

## Orbital involvement in sinonasal disease

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**Purpose** To present a number of cases of sinonasal disease with secondary orbital involvement and to review these disorders in the light of previously published reports.

**Methods** A retrospective study of patients with primary sinonasal tract disease and secondary orbital involvement who were admitted to the King Abdul Aziz University Hospital in Riyadh, Saudi Arabia from 1988 to 1993. Patients with disease causing concomitant involvement of the sinonasal tract and the orbit which did not originate in the sinuses were excluded, as were ambulatory patients.

**Results** The final study group comprised 28 patients (19 males and 9 females, ranging in age from 18 months to 65 years, mean 33 years) with the following diagnoses: nine bacterial sinusitis, eight fungal sinusitis, two mucocoeles, two osteomata, one fibrous dysplasia, one angiofibroma, one fibromatosis, and four carcinomas.

**Conclusions** Diseases of the sinonasal tract with orbital extension must be considered whenever a patient presents with signs and symptoms of orbital disease. Cooperation between the ophthalmologist and the otolaryngologist is desirable for proper management. *Saudi J Ophthalmol 1995;9:29-37.*

### Key words

orbital diseases – paranasal sinus diseases – paranasal sinus neoplasms – sinusitis

THE CLOSE RELATIONSHIP BETWEEN diseases of the sinonasal tract and the orbit is based on their anatomic proximity. The orbit is related superiorly to the floor of the frontal sinus, medially to the lateral wall of the ethmoid labyrinth, inferiorly to the roof of the maxillary sinus and posteromedially to the anterolateral wall of the sphenoid sinus. Some 60% to 80% of the orbital wall is made of sinus walls.

Concomitant orbital and paranasal involvements may occur due to lesions originating in the sinonasal tract or abnormalities that involve the sinuses and the orbit simultaneously, such as developmental or traumatic lesions.

The subject of this report is only those lesions that originate in the sinonasal tract. The purpose of the paper is to present a number of sinonasal disorders involving the orbit secondarily that were treated at the King Abdul Aziz University Hospital in Riyadh from 1988 to 1993 and to review these disorders in the light of previously published reports.

### Patients and Methods

A computer search was undertaken of patient records at the King Abdul Aziz University Hospital in Riyadh, Saudi Arabia to retrieve cases of primary sinonasal tract disease with secondary involvement of the orbit. Patients selected for the study had been admitted to the hospital in the five-year period 1988 to 1993.

The records were rejected of patients with diseases that caused concomitant involvement of

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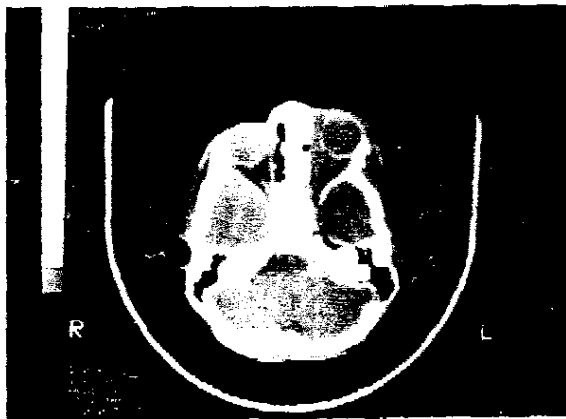
the sinonasal tract and the orbit but did not originate in the sinuses; examples of these were development anomalies, traumatic lesions, systemic diseases (such as *Wegener's granulomatosis*) and metastases. Patients treated in the outpatient clinic as ambulatory cases (such as those with preseptal edema) were also rejected.

Data concerning the age, sex, diagnosis, the sinuses affected, and the features of orbital involvement were retrieved from the selected records and collated to provide a description of a number of sinonasal diseases eventuating in orbital involvement.

### Results

Orbital involvement was demonstrated in 28 patients with primary sinonasal disease. In most patients, the sinuses and the orbital involvement were demonstrated radiologically. Histological and/or bacteriological examination substantiated the diagnosis in all cases, except the case of angiofibroma which was diagnosed by angiography. The distribution of the patients according to the diagnosis, age and sex is shown in Table 1.

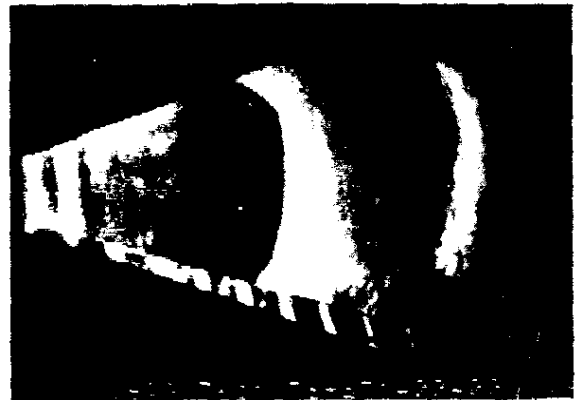
The nine patients with orbital infections (Figures 1, 2a, 2b, and 3 refer to some of these patients) presented initially to the ophthalmologist (two of these patients have been reported before?).



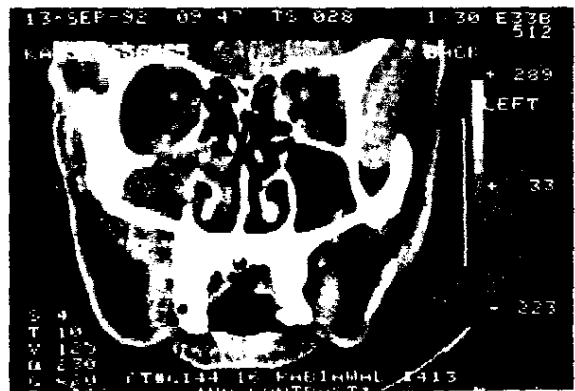
**Figure 1.** The axial CT scan shows a slightly enhancing soft tissue density in the ethmoid sinus and the medial orbital wall which is displacing the medial rectus muscle and causing mild proptosis. The orbital swelling may be due to inflammatory edema or a medial subperiosteal abscess.



**Figure 2a.** Right superior subperiosteal abscess. The axial CT scan shows retrobulbar soft tissue swelling with hypodense areas suggesting pus formation. The anterior and middle ethmoids are opacified.



**Figure 2b.** The ultrasound (B mode) of the same patient shows a hypoechoic area in the retrobulbar region suggesting abscess formation.



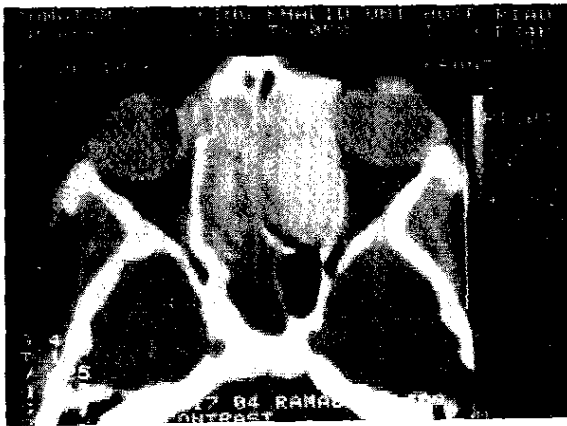
**Figure 3.** Right inferior subperiosteal abscess. The coronal CT shows a soft tissue mass in the maxillary sinus and orbital floor.

**Table 1.** Distribution of patients according to diagnosis, sex, and age, showing breakdown of tumors and tumor-like lesions.

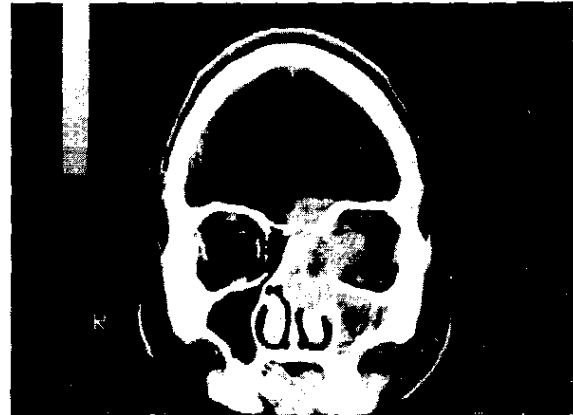
Diagnosis	Number	Sex		Age range (years)
		M	F	
Orbital bacterial infection	9	6	3	1.5-65
Fungal (aspergillosis) sinusitis	8	2	6	11-38
Tumors and tumor-like lesions	11	11	0	12-62
<i>mucocele</i>	2	2	0	22-29
<i>osteoma</i>	2	2	0	26-32
<i>fibrous dysplasia</i>	1	1	0	18
<i>angiofibroma</i>	1	1	0	12
<i>fibromatosis</i>	1	1	0	51
<i>carcinoma</i>	4	4	0	46-62
Total	28	19	9	

Eight patients had fungal (*Aspergillus*) sinusitis. Of these, one patient had the allergic type (Figure 4) and seven patients had invasive-type sino-orbital fungal disease (Figure 5). The maxillary and ethmoid sinuses were involved in all eight cases; the sphenoid was involved in four patients and the frontal sinus in only one. The patient with allergic-type aspergillosis developed bilateral mild proptosis (Figure 4) but had no visual deterioration. Of the seven patients with invasive-type sino-orbital aspergillosis, five had unilateral proptosis; this was accompanied by visual loss in two patients, one of whom presented initially to the ophthalmologist.

All the patients had surgical excision of the disease, followed by steroid therapy in the allergic patient, and by antifungal therapy in the others.



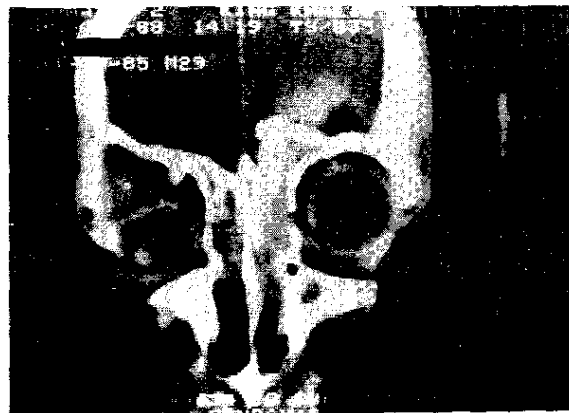
**Figure 4.** Allergic-type *Aspergillus* sinusitis. The CT scan shows non-homogenous opacifications of both ethmoid sinuses with bulging of the lateral ethmoidal walls causing mild proptosis.



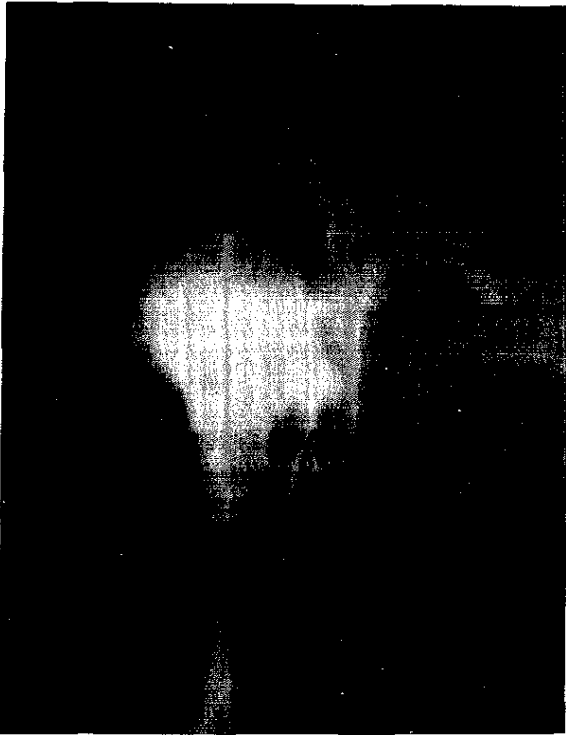
**Figure 5.** Invasive-type *Aspergillus* sinusitis. The coronal CT scan shows a heterogeneous soft tissue mass involving the left nasal cavity, maxillary sinus, ethmoid sinus, orbit and anterior cranial fossa.

Exenteration of the orbit was carried out in one of the patients with invasive-type sino-orbital aspergillosis who had complete visual loss but this failed to prevent the fatal outcome of the disease.

The remaining eleven patients in the series had tumors or tumor-like lesions. Two patients had mucoceles; the ethmoidal sinus was involved in one case, while the second patient had a mucocele in the fronto-ethmoidal sinus which had extended intracranially (Figure 6). Both patients presented initially to the eye clinic because of proptosis. Two patients had large fronto-ethmoidal osteomas (Figure 7) causing mild proptosis. Mild proptosis also affected the one patient with fibrous dysplasia

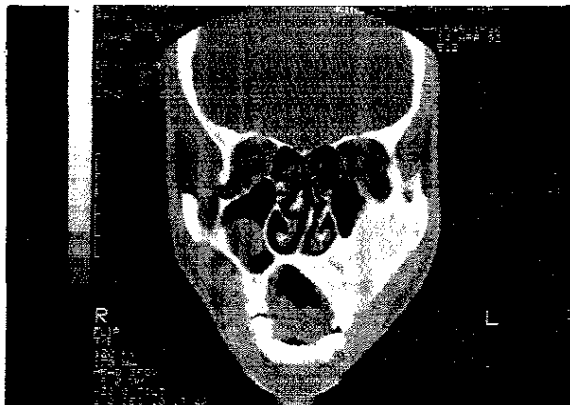


**Figure 6.** Mucocele (infected). The coronal CT scan shows a mass in the superior part of the right orbit which is displacing the globe downward and laterally. The cyst involves both frontal sinuses and there is intracranial extension on the left side.

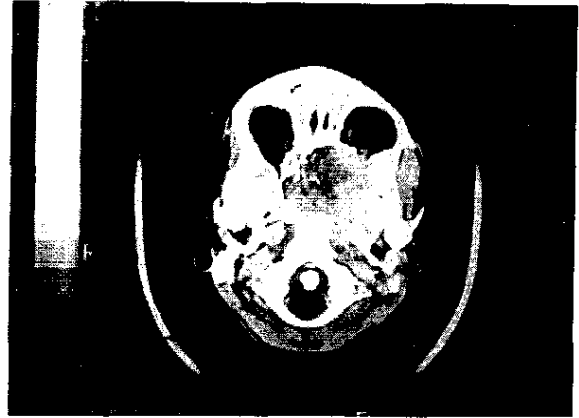


**Figure 7.** Osteoma. The tomogram displays a high-density, well defined mass involving the region of the right ethmoid and frontal sinuses.

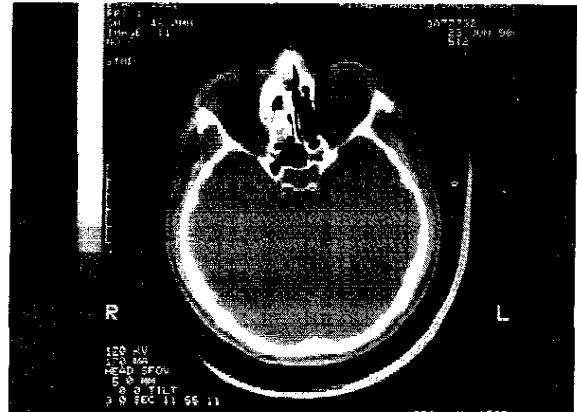
involving the maxilla, frontal and zygoma bones (Figure 8). The single angiofibroma case was diagnosed by CT, MRI and angiography (Figure 9); no biopsy was taken because of the risk of excessive bleeding. One case of fibromatosis, which has been documented previously,<sup>3</sup> involved the ethmoid, maxillary and frontal sinuses with extension to the orbit (Figure 10). Finally, four



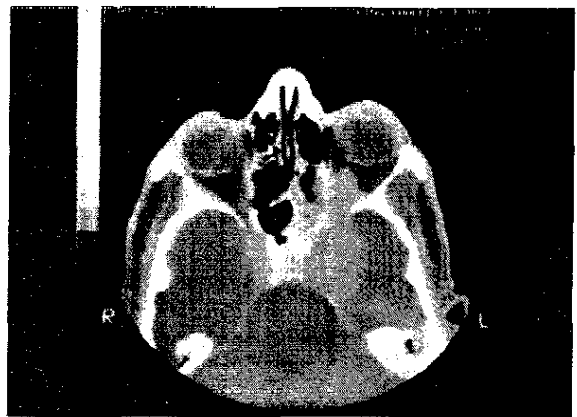
**Figure 8.** Fibrous dysplasia. A coronal CT scan showing a non-homogenous, high-density area involving the left maxilla.



**Figure 9.** Angiofibroma. This post-contrast CT scan shows a markedly enhancing soft-tissue mass at the skull base with encroachment on the posterior part of the left orbit.



**Figure 10.** Fibromatosis. An axial CT image showing a high-density mass involving the right ethmoid sinus with expansion of its lateral wall causing lateral displacement of the eyeball.



**Figure 11.** Carcinoma. An axial CT demonstrates a soft tissue mass involving the left posterior ethmoid sinus, as well as the orbit, parasellar area, and left temporal lobe region.

patients had paranasal sinus carcinoma with extensive orbital invasion causing proptosis and varying degrees of visual disturbance (Figure 11). The maxillary and ethmoid sinuses were affected in all the cases of carcinoma, while in one case the sphenoid was also involved.

### Discussion

Since this study group is small and comprises only selected patients, no attempt has been made to reach statistically significant epidemiological conclusions. The object of the study is to review a number of sinonasal diseases which secondarily involve the orbit. The treatment and prognosis of such disorders are not within the scope of this paper. However, a brief outline of the management of bacterial infection is presented to emphasize the importance of early aggressive therapy and of cooperation between the ophthalmologist and otolaryngologist.

#### Bacterial infection

According to the most commonly used classification system, orbital bacterial infections fall into two classes, each of which has a number of stages.<sup>4</sup>

- a) Preseptal infections occur anterior to the orbital septum and tarsal plate. There are three stages: edema, cellulitis, and abscess.
- b) Postseptal infections occur within the confines of the bony orbital walls and the eyelid septa. The five stages of postseptal infection are inflammatory edema, cellulitis, subperiosteal abscess, orbital abscess, and cavernous sinus thrombosis.

Sinusitis may give rise to the orbital apex syndrome; this is classified separately by some authors to include cases of optic neuritis.<sup>7</sup>

Orbital bacterial infection is reported to be the most common cause of unilateral proptosis in children; it is reported that 60% to 80% of such infections are secondary to paranasal sinusitis.<sup>4,6</sup> Acute sinusitis accounts for the preponderance of orbital infections in children, while in adults it usually occurs with exacerbation of chronic sinusitis.

This type of infection originates most frequently in the ethmoid sinus, followed by the

maxillary, frontal and sphenoid sinuses, in that order. In most instances, however, more than one sinus is affected at a time. In one study, 84% of patients were found to have infection in two or more sinuses, the ethmoid-maxillary combination being the most common.<sup>7</sup> Infection enters the orbit either by direct extension or by thrombophlebitis along the valveless venous channels. Direct extension is facilitated by the presence of thin bone or, less often, by congenital dehiscence of the bone between the orbit and the adjacent sinus.\*

The clinical features of orbital infection have been described in detail elsewhere.<sup>4-10</sup> Imaging evaluation of orbital infection is done by ultrasonography (Figure 2b), CT (Figures 1 and 2a), or MRI.

Ultrasonography is unreliable even in non-apical disease (in which bony walls interfere with the signal), although most treating physicians use this test to follow the response to medical and/or surgical therapy of disease initially imaged by CT or MRI scanning.

On CT scan, cellulitis manifests as a diffusely increased intraorbital soft tissue density. Early abscesses are difficult to differentiate from advanced cellulitis. Most practitioners will interpret a hypodense area in an inflamed orbit as an abscess. A subperiosteal abscess may appear as a displaced enhanced orbital periosteum adjacent to an opacified sinus. It is not easy to differentiate between a subperiosteal abscess and an orbital abscess. MRI is less useful because it does not visualize the bone, although it is preferred for evaluation of suspected intracranial complications.

Treatment is directed at both the cause of the sinonasal disease and the secondary orbital involvement, requiring full cooperation between the ophthalmologist and the rhinologist. Patients with preseptal infection may be treated with antibiotics, nasal decongestant and local heat application on an outpatient basis, although surgical drainage of the preseptal abscess and/or the infected sinus is sometimes needed.

Patients in whom postseptal infection is diagnosed should be hospitalized for immediate intensive therapy. Intravenous antibiotics that are effective against *Staphylococcus*, *Streptococcus*, *Hemophilus influenzae* and anaerobes and that also have the ability to penetrate cerebrospinal

fluid are essential. Serial ophthalmologic examinations (every 2 to 4 hours) to ascertain response to treatment are mandatory. If conservative treatment is successful, intravenous antibiotics are continued for 4 to 5 days after the patient's temperature returns to normal, and oral antibiotics are given thereafter for at least 4 weeks. Surgery is indicated when there is diminished visual acuity, evidence of subperiosteal abscess or more advanced disease, and failure to respond to medical treatment.<sup>4,8,9,10</sup> Drainage of the sinuses and subperiosteal abscess may be done endoscopically, intranasally or externally.<sup>8,10,11</sup> Drainage of an orbital abscess requires incisions in the periorbita.<sup>12</sup>

In cases of optic neuritis, the involved sinus should be drained immediately'; optic nerve decompression is required if signs of impaired vision do not begin to subside within 24 hours of sinus drainage or if the patient has substantially compromised vision at initial presentation.\*

### **Fungal sinusitis**

Aspergillosis is the most commonly encountered fungal infection of the sinuses. Of the eight patients in whom *Aspergillus* sinusitis was diagnosed, four patients were from Sudan where the disease is highly prevalent. There is some evidence, however, that the incidence of the disease in Saudi Arabia is actually higher than has previously been suspected.<sup>13</sup>

There are four clinicopathological types of aspergillosis of the sinonasal tract: allergic; non-invasive; invasive; and fulminant.<sup>14</sup> In allergic aspergillosis, the scanty fungal filaments in the lumen induce an allergic reaction in the mucosa. The disease may cause proptosis, especially in children, as a result of expansion (and sometimes erosion) of the sinus walls (Figure 4). Non-invasive aspergillosis (aspergilloma) is a localised disease confined in the sinus lumen and rarely involves the orbit. Invasive aspergillosis is characterized by a chronic granulomatous reaction in an immunocompetent patient; clinically and radiologically, it mimics a neoplasm of the sinus with invasion of the adjacent structures including the orbit (Figure 5). Fulminant aspergillosis is a rapidly progressive, destructive, necrotizing lesion affecting

immunocompromised patients; destruction of the sinus walls occurs rapidly with extension into the orbit and cranium.

Treatment of *Aspergillus* sinusitis entails extensive and repeated debridement of the disease, followed, in the allergic type, by steroid therapy and by adjuvant antifungal therapy in the invasive and fulminant types. Treatment should be aggressive, particularly in the fulminant type. If granulomatous fungal reaction encroaches upon the globe or if an orbital apex syndrome occurs, orbital exenteration is rarely avoidable. In this event, the defect should be allowed to granulate as a skin graft would add non-viable tissue for fungus propagation and might block early detection of advancing necrosis.<sup>4</sup>

The other fungal sinusitis known to involve the orbit (but not presented in this series) is mucormycosis (rhinocerebral phycomycosis). Seen mainly in poorly controlled diabetic patients, it is characterized by a progressive black necrotic lesion which extends rapidly from the nose to the orbit and cranium. Proptosis, early blindness, total ophthalmoplegia and lax, cold, non-tender periorbital puffiness have been described as typical signs of orbital ischemia and may provide the first indication of local pathology in many cases.<sup>15</sup>

### **Tumors and tumor-like lesions**

#### **Mucoceles**

Mucoceles are cysts lined by secretory sinus mucous membrane. They follow blockage of the natural sinus ostium. Mucoceles are most frequently found in the frontal sinus. As the frontal mucocele enlarges, it expands in the direction of least resistance-usually downward toward the roof of the orbit, causing the orbital contents to be displaced downward and laterally (Figure 6). Most ethmoidal mucoceles are part of extensive frontal-ethmoidal disease, although pure ethmoidal lesions are seen.<sup>16</sup> Ethmoidal mucoceles usually expand through the thin lamina papyracea, causing the orbital contents to be displaced laterally or downward.

Mucoceles of the sphenoid sinus may cause orbital apex syndrome. The maxillary sinus mucocele may cause upward displacement of the orbital contents, although may instead cause

enophthalmos due to loss of the antrum roof.

Most mucocele patients present to the ophthalmologist. In one study, proptosis was the most common presenting symptom in virtually all of the 98 patients seen with frontal-ethmoidal mucoceles.<sup>17</sup> Ocular motility is usually decreased in upgaze due to the presence of a mass in the upper inner quadrant which often has a characteristic "egg-shell" cracking sensation on palpation. Radiography of a mucocele demonstrates a smooth, globular lesion with punched-out areas of radiotranslucency.

Mucoceles are treated by complete surgical removal of the cyst and drainage of the sinus into the nose.

### *Osteomas*

Osteomas are slow-growing, benign tumors composed of a central core of cancellous bone surrounded by varying amounts of dense, compact bone.<sup>16</sup> They are most commonly found in the frontal sinus, the ethmoid sinus and the maxillary sinus, in that order. Their origin may be difficult to determine if they are large. In most instances they are asymptomatic and are diagnosed coincidentally during the work-up for unrelated complaints. They cause symptoms when they interfere with sinus drainage or, possibly, when they impinge on the orbit or the dura. Atallah and Jay (1981) reviewed 23 patients treated for paranasal sinus osteomas; ten of these patients presented with ocular symptoms and six of the ten had proptosis.<sup>18</sup> Osteomas are readily visualized by x-ray (Figure 7).

Symptomatic osteomas should be removed completely, while asymptomatic tumors should be followed up regularly as they may continue to grow.<sup>16</sup>

### *Fibrous dysplasia*

Fibrous dysplasia is a benign, self-limiting lesion which occurs almost exclusively during the first two decades of life. It is characterized by the replacement of normal bony architecture by a cellular fibrous tissue containing islands or trabeculae of metaplastic bone. Clinically, fibrous dysplasia may be confined to a single bone (monostotic) or may affect several bones (polyostotic). It occurs most often in the maxilla, the sphenoid bone or the frontal bone. Periorbital

swelling and asymmetry are common presenting signs of this lesion." Radiographs usually show a sclerotic lesion with a ground-glass appearance (Figure 8).

Small, stable lesions without significant cosmetic or functional deformity require observation only. The treatment for lesions causing either cosmetic or functional (visual or sinonasal) symptoms is generally accepted to be conservative excision.

### *Angiofibromas*

Uncommon and benign, but locally destructive, angiofibromas affect adolescent males almost exclusively. The tumor is composed of a rich vascular network in a fibrous stroma. The vascular channels are devoid of smooth muscles, contributing to the tumor's susceptibility to profuse bleeding. The tumor usually originates from the posterolateral part of the nose but extends readily to the pterygopalatine fossa. It may involve the orbit via the inferior orbital fissure (Figure 9). It may extend from the orbit to the middle cranial fossa through the superior orbital fissure. It is not unusual for the angiofibroma to extend into the orbit, but, fortunately, secondary attachment to orbital periosteum appears to be rare.<sup>16</sup> Approximately 10% of patients have proptosis at presentation.<sup>20</sup>

Angiofibromas should be removed through an approach tailored to the extent of the tumor. Embolization may be of value in reducing intraoperative blood loss. Excision of the tumor from the orbit can jeopardize vision and extraocular motility.\*

### *Fibromatosis*

Fibromatosis is a rare, histologically benign but clinically aggressive fibroblastic lesion arising from musculoaponeurotic structures throughout the body. The supraclavicular fossa is the most common site for head and neck fibromatosis but it has been reported to affect the nasopharyngeal tract and the orbit (Figure 10).<sup>3</sup> The tumor usually infiltrates the surrounding structures. Microscopically, the tumor is formed of well differentiated fibrous tissue which is usually highly cellular.

Complete excision of the tumor is the ideal treatment. If complete extirpation is not possible

due to involvement of important structures, radiotherapy and/or chemotherapy may be considered.

### *Carcinomas*

After Graves' orbitopathy and pseudotumors, secondary tumors from the paranasal sinuses are the third most common cause of proptosis in adults.<sup>4,6</sup> The majority of the tumors originate in the maxillary sinus and most of them are squamous cell carcinoma. The disease usually manifests at an advanced state, with orbital invasion (Figure 11) in almost two thirds of the patients.<sup>7</sup> Orbital involvement is generally considered to be an unfavorable sign, although its prognostic value depends on the site of involvement. Invasion commonly occurs through the lamina papyracea or through the infraorbital canal region, both of which are readily accessible to surgery. Of more sinister prognostic importance is invasion of the posterior medial wall from the posterior ethmoids with direct involvement of the optic nerve canal or infiltration of the retrobulbar structures from below, with spread in either case to the middle cranial fossa via the orbital apex.<sup>8</sup>

Treatment of sinonasal malignancy consists of a combination of surgery, radiotherapy and chemotherapy. Some authors have indicated that orbital exenteration improves survival for advanced operable disease, although analysis of other studies leaves this issue open to question.<sup>23</sup> At least 50% of patients eventually require orbital exenteration, while many treated by maxillectomy alone require support to the orbital floor.<sup>16</sup> However, given the observation that the tumor rarely penetrates the orbital periosteum, resection of the periosteum with preservation of the orbital content is advocated in cases where there is no involvement of the underlying orbital fat on frozen section.<sup>17</sup> The periosteum can often be grafted with a split skin graft. On the other hand, palliative removal of the eye is sometimes indicated to prevent painful proptosis.

### *Other tumors*

Many other tumors of the sinonasal tract, not presented in this series, have been reported to invade the orbit. Benign tumors include inverted papilloma, meningioma, schwannoma, and

neurofibroma. Malignant neoplasms include adenocarcinoma, mucoepidermoid tumor, adenoid cystic carcinoma, esthesio neuroblastoma, sarcoma, sinus glioblastoma, and ameloblastoma.<sup>20</sup>

### **Conclusion**

Diseases of the sinonasal tract with orbital extension must be considered whenever a patient presents with signs and symptoms of orbital disease such as orbital mass, proptosis, neurologic dysfunction of the eye, chemosis, or epiphora. Cooperation between the ophthalmologist and the otolaryngologist is clearly desirable for proper management of such lesions.

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