
SHORT REPORT

Paraganglioma of the cauda equina: case report and review of the literature

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Abstract
A rare paraganglioma of the cauda equina is reported. The clinical, radiological and histological features of this neoplasm are described. Diagnosis was resolved by the absence of glial fibrillary acid protein and electron microscopic evidence of neurosecretory features. Relevant literature is reviewed.

Key words: Cauda equina, computed tomography, electron microscopy, paraganglioma, retroperitoneal tumours, spinal tumour.

Introduction
Paraganglioma is the term used for the neoplasms that arise from the extra-adrenal paraganglion system—a collection of neurosecretory cell groups associated with the autonomic nervous system. The recognized paraganglia in man include those associated with: (1) some blood vessels and nerves in the head, neck and mediastinum; (2) the sympathetic ganglia; and (3) cell aggregates in various organs and peripheral vessels. While many of these bodies have been identified, many more exist along the craniosacral outflow of the autonomic nervous system and await discovery by astute investigators. More than half a century has elapsed since the awarding of the Nobel prize to C. Heymans for his discovery of the chemoreceptor nature of these bodies, and yet the basic issues regarding chemoreception remain unclear. Paragangliomas, rare neoplasms of this system, involve the head and neck region more frequently, particularly the carotid body and jugular paraganglia. However, they have been reported in numerous other sites as well. These tumours may be benign or malignant and functional or non-functional. A primary paraganglioma can occur in certain sites along the cerebrospinal axis. This paper gives a review of those tumours reported to primarily arise from the cauda equina. At least three hypotheses can be invoked to explain the primary occurrence of paraganglioma in the cauda equina region: it may arise from the paraganglial cells associated with the blood vessels; it may occur along the autonomic nerve fibres in the cauda equina; or the scattered ganglion cells known to be present in the fila terminales may provide the site for their origin.

Case report
A 28-year-old Saudi man was admitted with a 3½-year history of intermittent low back pain aggravated by sitting, standing and bending. The pain radiated to the posterior aspect of both legs. Increasing weakness and numbness...
of both legs started a year later. On admission he could only walk a few steps with help. Urinary retention requiring intermittent catheterization was present for many months prior to admission. He was initially treated by urologist and a cystoscopy revealed hypertrophic trabeculated bladder.

Examination revealed tenderness over the lumbar spine with marked weakness of flexors of both hips, as well as quadriceps, hamstrings and plantar and dorsiflexors of the feet. Pinprick sensibility was diminished over both the lower limbs. Knee and ankle jerks were bilaterally absent.

X-rays/tomograms of the lumbar spine (Fig. 1a, b) revealed deep scalloping and erosion of the bodies of the 3rd and 4th lumbar vertebrae as well as pedicular thinning and enormous widening of the lumbar canal at that level. Lumbar puncture failed to obtain any CSF and hence a C3C4 puncture was used to obtain a lumbar myelogram. This showed a large intradural defect causing a complete subarachnoid block with upper limit at the 2nd lumbar vertebra (Fig. 2a, b). Multiple, small circular filling defects in the dye column above this block indicated tumour seedlings. Spinal computerized tomography (CT) showed enormous widening of the lumbar canal with marked erosion of the bodies and neural arches of the 3rd and 4th lumbar vertebrae as well as extrusion of tumour nodules through enlarged intervertebral foramina into the retroperitoneal space (Fig. 3a, b, c).

Extensive laminectomy was performed from L1 to S1. Laminae of L3-4 were thinned and at places completely eroded by a friable mass that extruded into the back muscles. The dural tube was flimsy, enormously expanded and at places totally absent over a mass that extended from L2 down to S1. Large, ill-defined tumour nodules projected through thinned out and
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FIG. 2. Lumbar myelogram PA and LAT views showing: (a) complete intradural block opposite 2nd lumbar vertebra outlining crescentic upper pole of the tumour; (b) multiple small, circular filling defects above the block indicating tumour seedlings.

shredded dura and enlarged intervertebral foramina into the retroperitoneal space. Upon opening the already shredded dura a large (9.0 cm in greatest diameter) friable, soft, reddish-grey, poorly capsulated tumour mass was found to be filling a tremendously expanded lumbar canal. Some nerve roots were compressed and displaced laterally while others were enveloped by the tumour. No definite dural attachment of the mass could be seen. Multiple, small, greyish, grain-like deposits were seen on the nerve roots and arachnoid, both above and below the level of the tumour. With the help of magnification, microsurgical instruments and cavitron (ultrasonic aspirator) most of this vascular neoplasm, including the retroperitoneal extension, was resected and removed piecemeal, leaving an enormous cavity due to the marked erosion and excavation of the surrounding bony structures (Fig. 3). Nerve roots had to be dissected free of the tumour, but one of the roots entering the tumour had to be sacrificed. Gross tumour removal was almost complete except for the subarachnoid seedlings mentioned before.

Post-operatively, leg weakness improved rapidly, although the neurogenic bladder was slower to resolve. Six weeks later, the patient was continent and could walk without help. A post-operative VMA was negative.

Pathological examination

Light microscopic examination revealed the tumour to be composed predominantly of chief (Type I) cells, arranged in compact cell nests (Fig. 4). Reticulin stain demonstrated a variety in size and pattern of cell nests (Zellballen). The chief cells were round to oval, with central nuclei. These cells had granular or homogeneous eosinophilic cytoplasm and were PAS negative. The Grimelius stain was negative, as was the glial fibrillary acid protein (GFAP)
stain done by immunoperoxidase technique. Peripherally the cell nests showed scanty sustentacular (Type II) cells.

Electron microscopic examination revealed mostly chief cells rich in mitochondria and the characteristic intracytoplasmic membrane-bound dense core neurosecretory granules (Fig. 5).

**Discussion**

Paragangliomas of the cauda equina have only been recognized since 1970.6 A total of seven cases were reported in the decade that followed,5,7-12. Paragangliomas have not been mentioned in large tabulations of spinal cord tumours, perhaps reflecting the fact that they are not commonly recognized in this unusual
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In one case, the correct diagnosis was made only retrospectively after examination of recurrence 9 years after the initial surgery.

In the past decade much interest and data has accumulated about the biochemistry, immunohistology, ultrastructure and molecular biology of extra-adrenal paragangliomas. Increasing awareness of the cauda equina paragangliomas has resulted in a corresponding increase in the diagnosis and the reported incidence of these neoplasms. It now appears that these tumours may be less rare than was previously presumed.

Ours is the first such case to be reported from Saudi Arabia. The patient showed certain atypical features including enormous expansion of the lumbar spinal canal with scalloping, erosion, and invasion of the vertebral bodies and neural arches. Extrusion of tumour nodules through the thinned out dura and expanded intervertebral foramina into the retroperitoneal space, as well as subarachnoid seedlings, were the unique features of this tumour, rarely seen in the previously reported cases. All but one of the previously reported cases were entirely intradural, and osseous invasion and retroperitoneal extension was reported in only one case, and that too after surgery and radiotherapy. Spread of this tumour by subarachnoid seedlings has not been reported before.

A review of the reported paragangliomas in the cauda equina region shows that most patients were middle-aged and the age range at the time of diagnosis was 29 to 71 years. Males were more commonly affected than females. The most common symptom was low back pain with a duration ranging from a few days to 15 years. Motor/sensory deficits in legs, uri-
nary incontinence and paraplegia was seen in that order of frequency. None of the cases had a clinically evident hormonal syndrome. Except in the few bigger tumours, spine X-rays were normal. All patients had some myelographic block. Most of the reports do not give spinal CT findings. The neuroradiologic findings of a paraganglioma in the vertebral canal was described for the first time in 1985. Our case illustrates the characteristic CT appearances in the larger of these tumours.

A study of the operative findings showed that most of these tumours were vascular, well encapsulated and could be totally removed. Except for one case all the tumours were entirely intradural and extramedullary. Sites of origin/attachment of the tumours include filum terminale, conus medullaris, nerve roots or a vascular pedicle. A study of the gross characteristics of the reported neoplasms shows that they are generally circumscribed, soft, reddish, and range in size from 1.5 to 10.0 cm in greatest dimension. Osseous invasion and retroperitoneal extension was rarely seen. The microscopic features of paragangliomas of the cauda equina are similar to those located elsewhere in the body. The typical pattern on light microscopy is that of lobules of tumour cells separated by a vascular stroma (Fig. 4). The characteristic ultrastructural feature is the presence of intracytoplasmic neurosecretory granules (Fig. 5). Biochemically these neoplasms have been reported to contain dopamine, norepinephrine and epinephrine. Absence of GFAP and presence of neurosecretory granules on electron microscopy helps to differentiate a paraganglioma from the ependymoma of the cauda equina.

The majority of reported cauda equina paragangliomas were benign and capsulated and responded to surgical resection. We conclude that whenever feasible, gross total

FIG. 5. Electron micrograph of the tumour showing chief cell with membrane-bound dense-core granules.
removal should be the goal of surgery. However, when subtotal resection is necessary due to local invasion or extension, or when there is a doubt about completeness of surgical removal of the tumour, irradiation seems mandatory. Sufficient experience and follow-up is not yet available to evaluate the response to radiotherapy, but initial reports indicate that radiotherapy does not guarantee prevention of recurrence or regrowth of the residual neoplasm. In reported cases there was no recurrence or evidence of distant metastasis when the tumours could be removed in their entirety.

An awareness of the possibility of a paraganglioma in the cauda equina region will result in an increased recognition of this entity in Saudi Arabia, as happened elsewhere.

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References