Paraganglioma of the filum terminale internum

Report of a case and review of the literature

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Summary. A case of paraganglioma of the filum terminale is presented where normal sympathetic ganglion cells were seen in conjunction with tumour cells in a well-encapsulated tumour, suggesting a possible origin from heterotopic sympathetic ganglion.

Key words: Paraganglioma – Filum terminale internum – Neuroectodermal derivatives – Glomus

Paragangliomas are tumours arising from neuroectodermal-derived extra-adrenal paraganglia or chemoreceptor bodies. The central nervous system involvement may occur by: (1) direct extension from primary tumours of the glomus jugulare or glomus tympanicum [7, 18]; (2) spinal cord or brain compression from metastatic deposits in bone [5, 23]; (3) primary involvement in the pineal gland [20], pituitary gland [1] or filum terminale as well as the cauda equina [2-4, 8-16, 19, 21, 22]. Though metastases from malignant glomus tumours to other organs have been reported, no such occurrence in the central nervous system has been noted [6].

Our case of primary tumour of the filum terminale included normal sympathetic ganglionic tissue within a well-encapsulated tumour.

Case report

A 35-year-old man was admitted with low back pain of 2 years’ duration. The pain radiated down both lower extremities, was aggravated by strain and was worse at night. There was no history of trauma. For 2 months prior to admission he had numbness of the right leg and terminal dribbling upon urination.

Examination was normal except for flattening of the lumbar curvature, paravertebral spasm, limitation of flexion by pain, diminished patellar reflexes, hypoesthesia of the right saddle area and bilateral positive straight leg raising test. The complete blood count, urine analysis, radiographs of chest and lumbosacral spine were all normal. Lumbar myelogram disclosed a complete block of the contrast at the lower border of the L2 vertebral body with the appearance of an intradural tumour (Fig. 1A). A cisternal myelogram revealed the upper border of the block at the level of the mid L1 vertebral body (Fig. 1B).

With the diagnosis of a tumour of the cauda equina, laminectomy of L1-L2 was performed. Upon opening the dura a sausage-shaped purplish-grey mass was noted ventrolaterally within the cauda equina roots. The mass at its mid portion, was attached to the filum. Block resection of the tumour was achieved after clipping the filum terminale.

Postoperatively, there was full recovery and the patient was discharged without significant deficit.

Pathology. The mass measured 4 × 1.5 × 1 cm, with a fibrous band (part of the filum terminale) attached to its hilum. Its surface was smooth and glistening. The cut surface was dark red with scattered small grey areas, and was smooth and homogeneously soft.

Microscopically, with haematoxylin and eosin stain, the tumour had a well-defined fibrous capsule. The tumour cells were cuboidal to polygonal with cytoplasm resembling type-2 chief cells normally present in the chemoreceptor organs (Fig. 2). A few scattered cell nests with pale cytoplasm were also present. The tumour cells were closely packed in cords and nests with a fine organoid or alveolar pattern, well demonstrated in the reticulin preparation. In rare areas, the tumour cells were columnar with a pseudopapillary arrangement resembling the pattern of an ependymoma. No mitotic figures, cellular atypia or necrosis were seen. Subcapsularly, there was a cluster of ganglion cells with abundant pale cytoplasm and large vesicular nuclei containing prominent nucleoli intermingled with a few larger thick-walled blood vessels. Each ganglion cell was surrounded by cellular elements resembling capsule cells (Fig. 3). Phosphotungstic acid-haematoxylin stain for glial fibres and blepharoblasts, Masson-Fontana stain for argentaffin granules, Gomori’s reaction for chromaffin tissue and the PAS-McMauus method for glycosgen and mucin were all negative.

Electron microscopy. Tissue previously fixed in formalin was retreated for electron microscopy and examined with a Philips 400 electron microscope. The tumour cells were intricately interdigitated. Both light and dark cells were present. They had round to slightly ovoid nuclei with fine and coarse chromatin clumps that were more condensed beneath the nuclear membrane and a cytoplasm containing organelles and secretory granules. The light cells had more abundant and less dense cytoplasm (Fig. 4). The secretory granules characteristically revealed a dense central core separated by a clear halo from an outer single membrane (Fig. 5).
Discussion

Paraganglioma is a rare tumour found in the region of the filum terminale. Reviewing the available literature, we found 23 cases of paraganglioma of the spinal canal. Of these, 3 were epidurally located at the thoracic level and 21 were intradural, all at the level of the filum terminale and cauda equina (Table 1). The size of the tumours varied between 1.5 and 10 cm in maximum diameter with an average size of 4 cm. In 16 of the intradural cases a well-defined intact capsule was noted and 2 others were only partly encapsulated. The clinical presentation of this lesion is similar to that of other space occupying lesions of the cauda equina.

The histological pattern was predominantly alveolar, that is the tumour composed of organoid nests and cords of tumour cells. In 6 out of 24 cases pseudorosette formation suggested