Intrasacral myxopapillary ependymoma

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Abstract. Intrasacral ependymomas are rare, accounting for only a small fraction of primary sacral tumors. They are typically large at diagnosis, which is preceded by a long history of pain. We present a case discovered during investigation of infertility. MRI features of the myxopapillary subgroup are described.

Key words: Sacral neoplasm – Ependymoma – MRI

Ependymomas represent 60% of spinal cord gliomas [1] and 90% of those arising in the conus medullaris and filum terminale [1-4]. Most are myxopapillary, a distinct histologically recognizable subgroup so named by Kernohan in 1932 [5]. We report such a lesion arising in an unusual location in a patient who presented with infertility.

Case report

A 33-year-old man referred himself to the urology service complaining of a 10-year history of “infertility”. The initial physical examination was said to be normal. Laboratory investigations, including testosterone and follicle stimulating hormone were normal. Upon closer questioning, the patient admitted to ejaculatory and stress urinary incontinence. A semen specimen contained urine but the volume was otherwise normal. Decreased sperm count and motility were questionably related to small, bilateral varicoceles seen on ultrasonography. Antisperm antibodies were negative. Postejaculatory urinalysis was positive for sperm, indicating retrograde ejaculation. The patient was referred for cystometry which revealed delayed sensation to 325 cc, stress incontinence, and an incompetent bladder neck. It was concluded that retrograde ejaculation, the precise site of origin of the tumor may be obscure, and rare extramedullary spinal tumors such as ependymoma must therefore be considered. That most tumors of the filum terminale originate from the ependyma can probably be attributed to the number of ependymal cells within the filum terminale internus [8, 9]. Although ependymomas of other histologic types readily occur in the cauda equina/filum terminale region, the myxopapillary subgroup predominates [2, 3, 4, 8]. This tumor is characterized by well-defined cuboidal or columnar cells, arranged in a papillary fashion, with central cores composed of blood vessels and acellular hyaline connective tissue, which often stains for mucin [8]. Of 77 myxopapillary ependymomas reviewed by Sonneland et al. [4], 50 (65%) arose purely from the filum terminale internus [8, 9]. Although ependymomas of the conus medullaris and 4 (5%) were supracaudal. Intradural myxopapillary ependymomas seldom involve the sacrum [10]. These tumors are generally slow-growing and present in the third and fourth decades [2, 4, 8]. Although bone erosion or scalloping may be observed, gross destruction is rare [1, 10].

Although far less common, extradural myxopapillary ependymomas have been reported. Subcutaneous sacrococcygeal myxopapillary ependymomas occur in the in-
Fig. 1. a Anteroposterior plain film shows sharply demarcated lytic destruction in the sacrum (arrows). b Sagittal T1-weighted (600/15) MRI shows large, low signal, solid mass replacing upper sacrum (arrows). Note tongue of tumor extending into the lower lumbar canal (arrowheads). c Sagittal T2-weighted (2000/60) contrast-enhanced MRI shows heterogeneous hyperintensity of lesion. Note "pseudolayering" of contrast medium bladder (arrow). d Axial T1-weighted (700/22) contrast-enhanced MRI shows bright, heterogeneous contrast enhancement. e The tumor consists of papillary structures made up of ependymal cells (arrowhead) surrounding central cores containing blood vessels (arrow), hyaline acellular material (asterisk), and mucin (cross), characteristic of myxopapillary ependymoma. Hematoxylin & cosin, 100×

Myxopapillary ependymoma grows slowly and is therefore often large at diagnosis; like many sacral tu-