

Sacrococcygeal extraspinal ependymoma: a case report

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SUMMARY: Akpolat N, Bozlak N, Kazez A, Köseoğulları AA. Sacrococcygeal extraspinal ependymoma: a case report. Turk J Pediatr 2003; 45: 276-279.

Ependymomas, the common glial tumors of the spinal cord, occur occasionally outside the central nervous system and are called extraspinal ependymomas (ESE). ESE, which are clinically confused with other sacrococcygeal tumors, are rarely seen and found primarily in the sacrococcygeal region during childhood. We report a case of a seven-year-old boy presenting with a midline mass (6 cm diameter) over his coccyx. The solid mass was diagnosed as maxillopapillary type of ependymoma. Clinical and histopathological features of the case are described and literature reviewed.

Key words: extraspinal ependymoma, subcutaneous sacrococcygeal tumor.

Ependymomas usually arise within the spinal cord or in the brain. They account for 60% of tumors of glial origin in the spinal cord and comprise 90% of primary tumors in the filum terminale and cauda equina. Ependymomas rarely arise in the sacrococcygeal region (SCR) or other extraspinal sites. Sacral ependymomas, which are known as extraspinal ependymomas (ESE), account for less than 5% of all primary sacrococcygeal (SC) malignancies^{1,2}. As primary tumors, ESEs are found in subcutaneous tissues of the SCR and presacral space¹⁻⁴.

Case Report

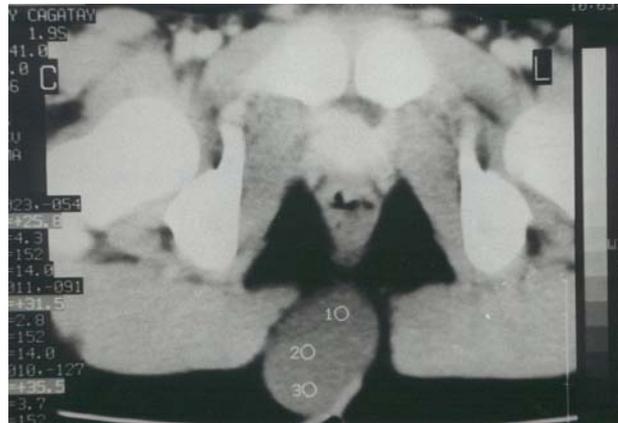
A seven-year-old boy presented with a midline mass over his coccyx, which had been present for six months. His parents did not notice any increase in the size of the mass, but pointed out

a hyperemic change during the last two months. He had no urinary or fecal incontinence.

A firm, subcutaneous, mobile, painless mass (6x5 cm) was palpated in the SCR. No presacral extension of the mass was felt during rectal examination. Neurological examination and pelvic and lumbosacral radiographs were normal. In computerized tomography, a solid, regular contour, homogeneous mass, localized in the extraspinal region, was observed and described as SC dermoid cyst (Fig. 1). The patient was operated based on the initial diagnosis of SC dermoid cyst, and a 6x5x3 cm nodular, well-defined mass was excised.

The histopathologic findings showed a mass of florid papillary structures composed of cellular connective tissue cores with central blood vessels

Fig. 1. Computerized tomography of the pelvis shows a solid sacrococcygeal mass.



(Fig. 2). In some regions, vascular structures were covered by a mucinous material. Most of the cells were cuboidal without notable pleomorphism, and rare mitotic figures were observed. The tumor was pseudoencapsulated with adjacent small nodules. Immunohistochemical examination findings showed extensive cytoplasmic glial fibrillary acidic protein (GFAP) (Fig. 3) reaction and vimentin positivity, whereas keratin and S-100 protein were negative. The histopathologic and immunohistochemical features of the tumor were identical with myxopapillary ependymoma.

The postoperative period was uneventful. During 21 months follow-up, the patient is alive, with no local recurrence, invasion to adjacent tissue

or distant metastasis of the tumor.

Discussion

In 1902, Mallorys cited the first case of an ESE. As of 2000, 22 pediatric SC ESEs have been published in the literature². The most common site for primary ESEs is in the subcutaneous SC and presacral region^{1,3}. In 1979, the World Health Organization classified ependymomas into three subtypes, including myxopapillary papillary and subependymal. The majority of sacral ependymomas are myxopapillary type^{1,4}, as in the present case.

Helwing and Stern⁷ discussed four situations of involvement outside the central nervous system (CNS) for ependymomas: 1) Metastasis

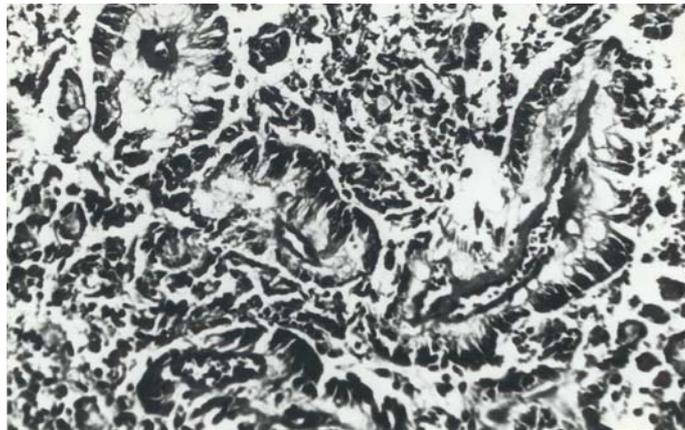


Fig. 2. The tumor consists of papillary projections, pseudorosette formation, and fibrovascular core surrounded by cuboidal cells (hematoxylin and eosin H&E, x100).

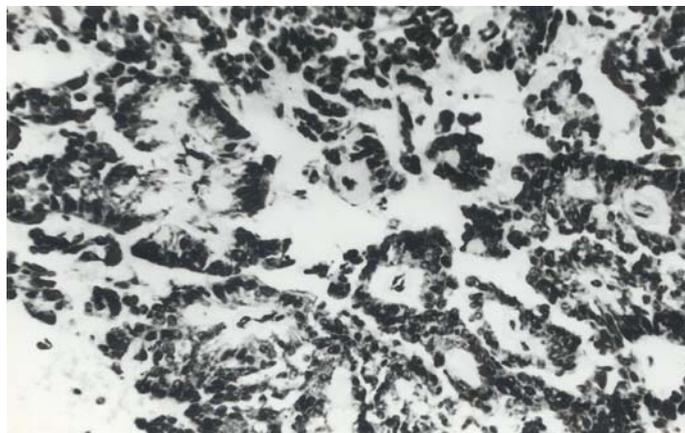


Fig. 3. Glial fibrillary acidic protein (GFAP) immunoreactivity is present within the tumor cells (GFAP, x200).

or direct extension following surgical excision of a primary tumor from the CNS, 2) Direct extension to the soft tissues of the SCR from a primary ependymoma of the lower spinal cord, cauda equina or filum terminale, 3) Occurrence as a primary tumor of the skin and, 4) Subcutaneous tissues of the SCR without demonstrable connection with the spinal cord. According to this classification, the present case is in the last group.

The origin of ESEs is still unclear. The embryologic basis for the origin of these tumors seems to be from coccygeal medullary vestige³. By the end of the third month of gestation, only a remnant of the caudal portion of the neural tube remains at the tip of the coccyx. This ependymoma-linked cavity appears to have pinched off from the neural tube and assumes an extramedullary and, at times, even an extraspinal position^{1,3}. It is seen not only in embryos at the site of closure of the posterior neuropore (postanal pit), but according to Bale⁹, also in over 50% of infants up to the age of one year¹. The coccygeal medullary vestige, therefore, may be the source of heterotopic ependymal cell rests^{2,3}. Presence of such developmental failure demonstrates incomplete closure of the neural arch, enhancing the possibility of ependymal heterotopia. Presacral ependymomas are postulated to arise either from the cauda equina, extending through the sacral foramina into the soft tissues anteriorly, or from extradural remnants of the filum terminale^{3-7,10-13}.

Subcutaneous myxopapillary ependymomas are also rare. The female: male ratio is nearly equal. The age of patients with symptoms varies from 10 months to 47 years. Ependymomas arising in the CNS, cauda equina and presacral region are associated with neural dysfunction, whereas these symptoms are absent in purely subcutaneous myxopapillary ependymomas. A slowly progressing, sometimes-painful mass in the intergluteal fold is often misdiagnosed preoperatively as a pilonidal cyst, teratoma, sweat gland tumor or chordoma^{8,14-16}.

Histologically, ependymomas arising in the SCR are mostly of the myxopapillary type, which resemble those arising from the filum terminale and cauda equina. Myxopapillary ependymoma generally contains a papillary configuration covered by cuboidal or short columnar cells. The perivascular stroma shows a characteristic mucoid

or myxomatous degeneration^{8,16,17}. The present case was myxopapillary type of ependymoma and had typical histopathologic features. Morpho-logically, the tumor did not show features of teratoma or of other tumors with which it's commonly confused. Sacrococcygeal cystic teratomas are histologically composed of three germ line elements, such as skin and cutaneous adnexal structures, cartilage, bone, bronchial or gastrointestinal epithelium, and mature glial tissue. In the present case, the tumor showed neither cystic components grossly and radiologically, nor features of teratoma histopathologically.

The treatment of subcutaneous SC ependymoma is wide local excision, if possible. Coccygectomy may be required, if the tumor is attached to the bone. There is local recurrence after 0.5 to 13.5 years in approximately 25% of patients after the initial surgery. Recurrences at the initial excision site often precede distant metastasis, which may develop in 17% to 20% of the patients^{1,2,18}. The common sites of metastasis include the inguinal lymph nodes, lungs, pleura, bones and liver^{1,3}. Up to 70% of patients die due to widespread metastasis¹⁸. In the present case, no recurrence, invasion to adjacent tissues or distant metastasis occurred during postoperative 21 months' follow-up.

Radiation has proven to be unsuccessful as a primary therapeutic option^{1,18}. However, adjuvant radiotherapy has been recommended as postoperative treatment when the initial resection was incomplete, after the removal of a local recurrence, or if metastases are present. Chemotherapy is reported to be of no benefit¹.

Based on our case and a review of the literature, we concluded that an extraspinal ependymoma is a form of tumor which may present as a mass in the subcutaneous soft tissue posterior to the SCR and is that it usually mistaken for a pilonidal cyst, lipoma or epidermoid cyst. It is advisable to consider a SC ependymoma as being a low-grade malignant tumor, and complete local excision should be performed in all cases. Since the tumor may recur locally and tends to metastasize, such patients should be examined periodically.

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