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Case study

Ependymomas arising outside of the central nervous system: A case series and literature review

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ABSTRACT

Background: Extracranial and extraspinal ependymomas are extremely rare tumors mostly published in the literature as case reports.
Methods: MD Anderson Cancer Center institutional database was screened for patients with extra CNS ependymomas over a 25 year period.
Results: Eight patients with extra CNS ependymoma were identified. Five cases originated in the sacrum or subcutaneous sacral area, and the other 3 cases originating in the breast, lung and adnexa of the uterus. By histology, most cases were myxopapillary ependymomas (5 cases), 2 cases were grade II ependymoma and 1 case was an anaplastic ependymoma. Metastases occurred in 6 cases and 3 patients died due to progressive disease. Most cases required surgery, radiation and chemotherapy.
Conclusion: Extra CNS ependymomas are very rare tumors that tend to metastasize, even though most cases are histologically classified as low grade ependymomas.

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1. Introduction

Ependymomas are rare neuroepithelial tumors originating from the wall of the ventricles or the central canal of the spinal cord and composed of neoplastic ependymal cells. They constitute only 1.9% of primary central nervous system tumors among adults, but are the most common spinal tumors in adults younger than 19 years of age [1]. Extra-cranial and extra-spinal ependymomas are extremely rare with most reports describing one or 2 cases per institution and tending to occur mainly in children and young adults [2–8]. Reported sites where ependymomas have been described to arise outside the CNS include the mediastinum, lung, ovary, broad ligament, and sacrococcygeal region, with the latter region being the most common amongst the listed sites [2–5,7–10]. We here present a case series of 8 cases of extra CNS ependymomas seen at a single institution over a 25-year period with a review of the literature.

2. Methods

We conducted a retrospective data review of all adult non-CNS ependymoma patients in the MD Anderson Cancer Center (MDACC) institutional database from 7/1989 through 07/2014 under a protocol with waiver of consent approved by the Institutional Review Board (PA12-0619). All patients were age 18 or greater and had undergone a biopsy or surgical resection with a pathological diagnosis of ependymoma. Patient's demographic and clinical characteristics, treatments and outcomes were reviewed. Death was confirmed by review of medical records and/or death certificate.

3. Results

3.1. Clinical description

Eight cases of extra CNS ependymoma were seen at MD Anderson over a 25 year period. Table 1 summarizes the more salient clinical information on these 8 patients. The median age at diagnosis was 37 (range 19–76), with the majority of cases occurring in

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women (6 out of 8 patients). Most arose in the sacral and sacrococcygeal region (5 out of 8 patients), with a slight female predominance (3 out of 5 patients). All ependymomas arising outside of this region occurred in females (Cases 1, 3, 4). By histology, the myxopapillary variant was the most common histological subtype. Distant metastases developed in 75% of cases throughout the course of the disease. Despite this, only 3 of our 8 patients have passed away after a median follow up of 6.5 years. Treatment with surgery, radiation and chemotherapy was applied in most cases. A gross total resection was achieved in only two of these cases at the time of presentation.

Below are 3 cases described in more detail that highlight some important clinical features of this rare tumor entity (Correspond to cases in Table 1)

3.2. Case 1

55-year-old woman who on a chest X-ray done as part of a routine physical examination, was found to have irregularly lobulated right-sided lung lesions. CT of the chest confirmed the presence of multiple right-sided pleural-based masses (Fig. 1A). Resection showed these lesions to be ependymoma, WHO grade II. The patient remained clinically and radiographically stable off treatment over a 6-year period, at which point imaging showed evidence of recurrence in the lungs. She underwent another resection with pathology confirming the diagnosis of ependymoma WHO grade II. Tumor cells were positive for estrogen (ER) and progesterone (PR) receptors. An immunostain for c-erbB-2/HER2/neu tyrosine kinase was negative. Once again she was closely monitored over time with evidence of slow growth occurring after 6 years. She was then started on tamoxifen since the tumor stained positive for ERs. She was stable over a period of 4 years, at which point she asked to discontinue tamoxifen. Imaging 6 months later showed evidence of radiographic progression with the primary lesion in the right lower lobe measuring 2.7 cm compared to 1.7 cm on prior scan 6 months earlier. The patient preferred to avoid a biopsy or resume tamoxifen, as long as she continued to be clinically asymptomatic.

3.3. Case 2

A 38-year-old gentleman presented with sacral pain and pain on defecation. MRI of the pelvis showed the presence of a large heterogeneously enhancing mass originating from S2 (Fig. 1B). Subtotal resection was consistent with a myxopapillary ependymoma WHO grade I (Fig. 2). Six months later, imaging showed evidence of tumor growth. He was treated with proton radiation and placed on surveillance. Two years later, staging revealed growth of the sacral mass, with pulmonary and soft tissue (gluteal) metastases. The patient was started on lapatinib and temozolomide. However, after 2 cycles, MRI showed an increase in the gluteal lesion for which he was treated with RT and started on cyclophosphamide and cisplatin. However, he had evidence of recurrent disease 4 years later and was started on etoposide for recurrent disease.

3.4. Case 3

A 19 year-old woman initially presented with hematuria with imaging revealing the presence of a mass within the bladder. At that time, she had also felt a right breast mass for several months. Breast excisional biopsy revealed the presence of myxopapillary ependymoma WHO grade I. Biopsy of the bladder mass also confirmed the diagnosis. Patient was treated with multiple chemotherapeutic agents including cyclophosphamide, adriamycin, etoposide, cisplatin, and procarbazine, but did not respond to any of these agents. She developed progressive disease involving the liver, lung, lumbar spine and subcutaneous tissue of chest wall, supraclavicular region and shoulder. The patient was treated with focal RT to the skin lesions, lumbar spine and embolization to the liver metastasis followed by paclitaxel for 3 cycles with some temporary stabilization of her disease. However, 6 months later she presented with hemorrhage and infarction in the left lobe of the liver from an enlarging metastatic lesion. In view of her rapid decline the patient was referred to hospice and passed away 5 years after initial diagnosis.

4. Discussion

Extra-CNS ependymomas are extremely rare tumors published mostly as case reports in the literature. Here we present 8 cases of extra-CNS ependymoma, which to the best of our knowledge is the largest case series published to date. Table 2 summarizes all cases of extra CNS ependymoma published in the last 25 years.

The most common location of ependymomas outside the CNS is in the subcutaneous sacrococcygeal or pre-sacral region, which was also the case in our patient series (5 out of 8 cases) [10]. It is assumed that the sacrum might be the most frequent location because of its proximity to the spinal cord in general and filum terminale in particular. While subcutaneous sacrococcygeal tumors are thought to arise from the coccygeal medullary vestige (an ependymal lined cavity forming the remnant of the caudal portion of the neural tube) the presacral tumors are thought to arise either from the extradural remnants of the filum terminale or as an extension from the intradural filum terminale [10,11]. Depending on the extent of disease, common presentations include pain, bowel/bladder dysfunction (as seen in case 3) as well as sacral root involvement [10]. Myxopapillary ependymoma is the more common histological type arising in this region (4 of the 5 tumors in our case series) [11].

Extra-spinal ependymomas arising outside of the sacrococcygeal or pre-sacral region are even rarer [2–6,9]. While histologically most of the sacral extraspinal ependymomas are a myxopapillary variant, tumors arising in extrasacral sites tend to exhibit more architectural variability than those in the CNS or in the sacral regions [12].

In this case series we describe ependymomas arising from breast, lung, and adnexa. While the sex distribution of sacrococcygeal tumors mirrors that of CNS ependymomas, extra-axial ependymomas arising elsewhere seem to arise only in women [5]. This pattern was also seen in our cohort. The pathogenesis of non-sacral extra-CNS ependymomas is unclear. Ependymal rests have not been described in the ovary, the uterine ligaments, or the mediastinum. One theory is that ependymomas in the ovary, broad ligament, and mediastinum might originate from neuro ectodermal teratoma tissue [13]. Another theory is that misdirected primordial germ cells transform to ependymal cells under the influence of female hormones, and this could explain the female predominance of such tumors [5]. It might also explain why extra-CNS ependymomas stain strongly and diffusely positive for estrogen and progesterone receptors [14]. The presence of hormonal receptors might explain the increase in size of the gluteal lesion that occurred during pregnancy in one of the cases presented (case 8). Additionally another patient (case 1) whose tumor cells were positive for ER and PRs was treated with tamoxifen with disease stability for 4 years. To our knowledge, this case is also the first description of an extra-axial ependymoma arising from breast tissue.

Extra CNS ependymomas, though typically, histologically classified as being of low malignant potential, these tumors are well known to have a higher propensity to metastasize. Surprisingly

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