Clinical Oncology: Case Reports

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Primary Subcutaneous Sacrococcygeal Ependymoma with Lung Metastasis: A Case Report

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Abstract

Ependymomas arising outside the central nervous system are rare. We here report a case of a 37 years old male not known to have any chronic medical illnesses apart from the history of recurrent pilonidal sinus that was treated surgically 5 years before his presentation. The patient presented with a painful enlarging sacrococcygeal area mass for 1 year. Imaging studies and tissue biopsy of the mass revealed sacrococcygeal maxilopapillary ependymoma with lung metastasis. Local excision of the primary mass was carried and 2 months after the primary procedure, thoracotomy for the possibility of metastasectomy was aborted due to disseminated. Local recurrence was evident 6 months after the primary procedure. The patient has received adjuvant chemotherapy for a total of 9 cycles and was on regular surveillance for a total of 11 months from starting the chemotherapy which showed stable disease.

Myxopapillary ependymoma despite its low grade-can metastasize and recur early. Wide local excision of the primary lesion is recommended even in metastatic disease. Role of adjuvant chemotherapy still questionable despite evidence of disease stabilization. Adjuvant radiotherapy to decrease local recurrence is an option that needs further research.

Keywords

Subcutaneous; Sacrococcygeal ependymoma; Lung metastasis

Introduction

Ependymomas are tumors that originally arise from glial cells that line cerebral ventricles and the spinal canal [1]. Most of these tumors are benign and slow-growing. Ependymomas typically develop within the Central Nervous System (CNS), very rarely ependymomas can develop outside the CNS. Sacrococcygeal ependymomas are thought to arise from a group of heterotopic ependymal cells that rest in this region as neural tube tissue remnant called coccygeal medullary vestige [2]. In the present case, we report a primary subcutaneous sacrococcygeal ependymoma with lung metastasis in a 37 years old male.

Case report

A 37-year-old Saudi male, not known to have any chronic medical illnesses apart from the history of recurrent pilonidal sinus, which was excised twice from the upper gluteal cleft 5 years before initial presentation to our center.

The patient started to complain of coccygeal mass that was first noticed one year before seeking medical advice. It was initially painless but became painful and enlarged gradually. He denied any incontinence, tenesmus or any gastrointestinal (GI) symptoms.

On examination, there was a single irregular mass in the gluteal area measuring around 10 × 8 cm with hyperpigmentation of the covering skin. The mass was round, firm, and immobile with a smooth surface. There was no tenderness or hotness. On digital rectal examination, the sphincter tone of the anal canal was suboptimum. The lumen of the anal canal and lower rectum showed no palpable masses, no ulceration, and no blood.

Patient workup included pelvic MRI, biopsy from the mass, CT chest abdomen, and pelvis, as well as, a lower GI Endoscopic Ultrasound (EUS). The Pelvic MRI (Figure 1) showed large necrotic nodular soft tissue mass measuring around 10.9 × 7.1 × 9.5 cm in transverse, anteroposterior and craniocaudal dimensions respectively. The mass was inseparable from the posterior anorectal canal deeply and extending to just below the skin superficially. The mass was reaching the tip of the coccyx and the gluteal muscles with no evidence of gross invasion seen. The mass showed heterogeneous T2 signal intensity and mostly low T1 signal intensity with evidence of peripheral enhancement and necrotic center.

The biopsy (Figures 2-4) showed papillary architecture with true fibrovascular cores in an abundant myxoid background. The tumor cells show pseudorosette pattern formed of neoplastic cells nuclei arranged radially around the vascular structure. The tumor cells contained round nuclei with small nucleoli, coarse chromatin, and moderate to plump granular eosinophilic cytoplasm. Tumor cells showed moderate nuclear atypia and mitosis at a rate of 15 MF/10

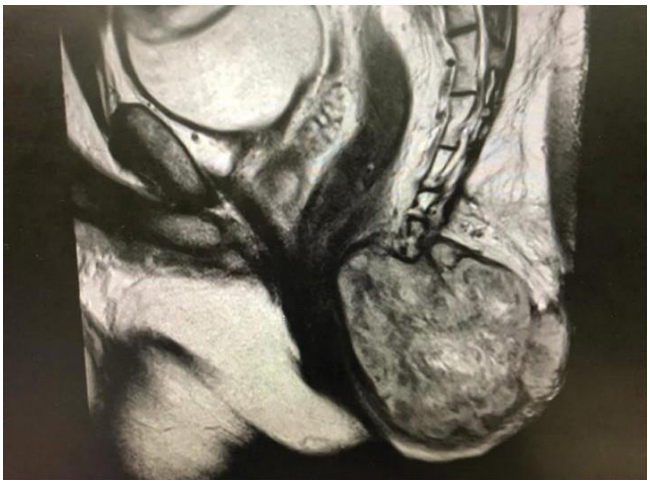

Figure 1: Sagittal view of T2-weighted MRI showing a nodular soft tissue mass extending from the skin superficially and deeply inseparable from the posterior anorectal canal.

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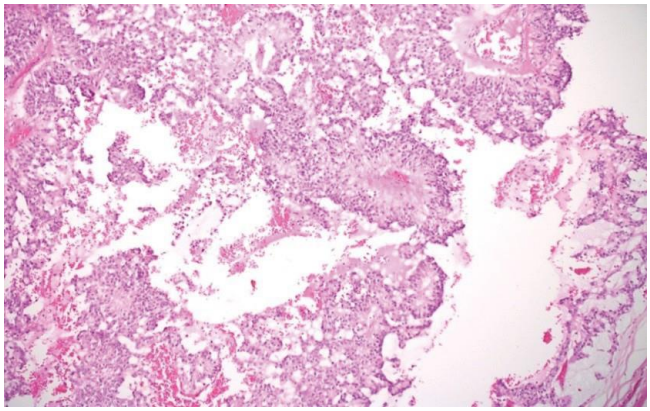

Figure 2: Papillary and perivascular pseudorosette architecture composed of polygonal cells with round nuclei showing mild pleomorphism, moderate eosinphilic granular cytoplasm, and rare mitosis. The background is myxoid Hematoxylin and Eosin stain, x200 magnification.

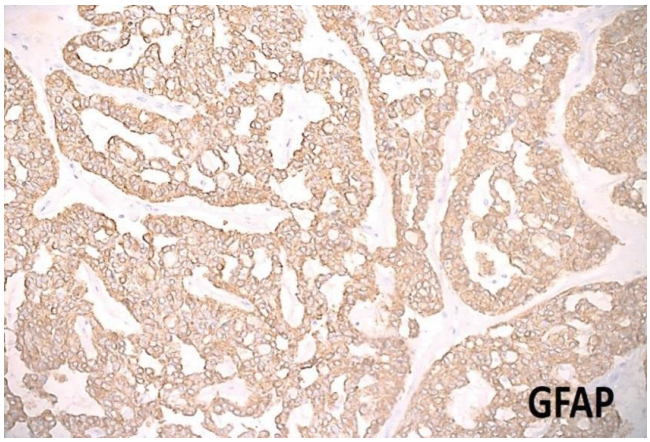

Figure 3: Tumor cells show diffuse membranous and cytoplasmic positivity for Glial Fibrillary Acidic Protein (GFAP) immunohistochemistry, x200 magnification.

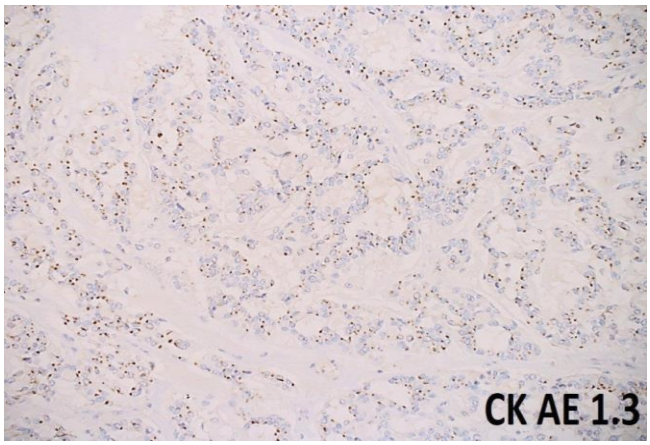

Figure 4: Tumor cells show diffuse perinuclear dot (golgi-pattern) positivity for Cytokeratin AE 1.3 immunohistochemistry, x200 magnification.

hpf. By immunohistochemistry, tumor cells were diffusely positive for GFAP and S100, both cytoplasmic and membranous; and CK/PAN showed focal perinuclear dot-like positivity (Golgi-pattern). While Ber-ep4, HBME1, EMA, CK7, CK20, and CEA were negative. Ki-67 was positive in 30% of tumor cells nuclei. The final diagnosis was myxopapillary ependymoma with anaplastic features.

Chest CT scan showed multiple pulmonary and pleural based nodules likely metastatic, the largest was in the right lower lobe measuring about 2.6 × 2.4 cm.

Further investigation included a CT-guided lung nodules biopsy, EUS, and bone scan. The tru-cut biopsy from the lung nodule confirmed metastasis. The histomorphology of tumor cells were similar to those in the specimen obtained from the sacrococcygeal mass. Bone scan was negative for metastatic involvement. Rectal EUS (Figure 5) showed that the tumor was not grossly invading the anal sphincters.

Tumour board and multidisciplinary team meeting were in favor of complete oncological resection of the primary tumor followed by chemotherapy and close follow up for possible metastasectomy of the largest lung nodules after chemotherapy response assessment.

The patient refused any option of non-sphincter preserving surgery despite the higher risk of recurrence expected.

Based on the patient's preference, wide local excision of the mass with limited margins excision near the anal sphincter (aiming not to damage the stretched sphincter) was performed. Intraoperatively, the tumor looked very well encapsulated, the surrounding muscle fibres were stretched but not grossly invaded. The mass was excised completely. Majority of the resection was with the use of the electrocautery as sharp dissection, while the bipolar device was used to control some of the feeding vessels.

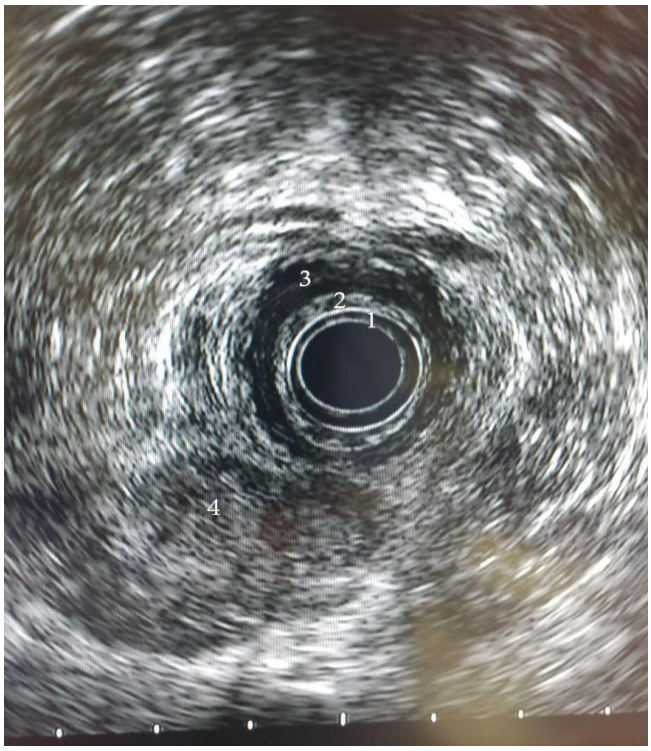

Figure 5: Rectal EUS revealing the tumor not invading the anal sphincter [1], endoscopic probe [2], internal anal sphincter [3], external anal sphincter [4] tumor.

The final histopathology report was consistent with myxopapillary ependymoma with anaplastic features (WHO grade 3).

The post-operative course went smooth and the patient was assessed by the medical and radiation oncology as well as Thoracic surgery. The patient denied any incontinence post-op.

Two months after the primary procedure, the patient underwent right exploratory thoracotomy for the possibility of metastasectomy. The disease was disseminated involving the pleural cavity, anterior chest wall, diaphragm and over the pericardium. Frozen section biopsy was consistent with the disease and the procedure was aborted.

On the follow-up imaging after 6 months the patient was found to have local recurrence measuring 3.5 × 2.9 × 2.5 cm. a multidisciplinary team meeting was held and the patient was started on single-agent cisplatin chemotherapy and received 5 cycles.

After completion of 5 cycles, the patient underwent restaging CT chest, abdomen and pelvis. Which showed stable disease and no progression. The chemotherapy regimen was changed to carboplatin AUC 6 and he received 4 cycles of treatment.

The patient is on regular surveillance and last imaging showed stable disease and no progression for a period of 17 months from the time of the primary procedure.

Discussion

Extradural ependymomas are extremely rare. The first case of extradural sacrococcygeal subcutaneous ependymoma was reported by Mallory in 1902 [3]. In more than a century, approximately 90 cases of extradural ependymomas were reported in English-language literature [4]. This type of tumors usually affects children younger than 8 years old and adults in their fourth decade, however, cases of 2 months old children and up to 67-year-old adults were reported. Gender predilection is skewed towards male gender [5].

Ependymomas are classified based on their location or histology. Based on their location, they are classified as CNS, sacrococcygeal, pelvic or extrapelvic [6]. Sacrococcygeal ependymomas are the commonest among them and usually they develop in the presacral soft tissue or in the subcutaneous tissue of this region. Pelvic and extra pelvic ependymomas exclusively affect females, mainly in their childbearing age. Based on histology, ependymomas have 3 subtypes: 1) myxopapillary ependymoma: 2) “classic” ependymoma and 3) anaplastic ependymoma. WHO regarded the first subtype as grade 1 and the second and third subtypes as grade 2 and 3 respectively; however, there is no clear cut association between the grading system and disease prognosis [7]. Most subcutaneous sacrococcygeal ependymomas are of myxopapillary histological subtype [5], and they have been reported to develop in ovaries, the uterine ligament and the mediastinum [1].

Subcutaneous sacrococcygeal ependymomas are commonly misdiagnosed as pilonidal disease because of their similar presentation near the gluteal cleft [8]. We believe that our patient was most probably misdiagnosed as pilonidal disease. Other differential diagnosis includes lipoma, teratoma, chordoma, myxoid chondrosarcoma, metastatic carcinoid and metastatic mucoid carcinoma [9]. CNS ependymomas rarely metastasize outside the CNS and extradural ependymomas rarely metastasize into the CNS [1]. However, extradural ependymomas carry a higher risk of systemic metastasis compared to CNS ependymomas that originate in the cauda [10]. The most common metastasis site of myxopapillary ependymomas in the lungs, followed by pleura, thoracic and abdominal lymph nodes and lastly the liver [1].

Recommended treatment option for subcutaneous sacrococcygeal ependymomas is wide local excision of the tumor with negative margins [8]. The proximity of the lesion in our case to the anal sphincter made the surgical planning a challenge. The possibility of sphincter involvement based on imaging made the option of excising the entire anal canal (abdominoperineal resection) a better oncological resection option in our opinion, but the patient refused that option. There haven’t been similar cases reported or clear guidelines with such close relationship between the tumor and the anal sphincter to support a negative margin only versus a more radical resection (entire resection of the anal canal and permanent stoma). Our surgical plan was more palliative based on the decision of the tumor board and the discussion with the patient. However, the intra-operative clear plane was assuring and the final pathology confirmed that there was no sphincter invasion. The literature supports coccygectomy if the tumor is attached to the coccyx, with the addition of adjuvant radiotherapy [11].

Chemotherapy was found to have no role in the treatment of primary non-metastatic non-recurrent myxopapillary ependymomas [8]. Due to the lack of studies and low incidence of these tumors, there have been no guidelines for adjuvant chemotherapy or radiotherapy and its use was all based on previous reports [12]. Temozolomide (TMZ) was reported to have controversial effects [13], whereas platinum-based and nitrosourea chemotherapy was labeled to have minimum effect [14]. Sorafenib was used as a multikinase inhibitor in one case report and showed a one-year stabilization of metastatic disease but the patient developed a reversible neuropathy [12]. Although no strong evidence is present supporting chemotherapy use and its effect on disease progression and despite local recurrence in our case, we used platinum-based chemotherapy that seemed to stabilize the disease for 11 months after the start of chemotherapy.

Since metastasis can develop decades after the initial presentation, prolonged surveillance is highly recommended [8].

Conclusion

In summary, Sacroccygeal Ependymomas are the commonest location to exist outside the CNS. Myxopapillary ependymoma despite its low grade can metastasize and recur early.

Complete surgical resection is so far the best treatment option. Even in metastatic disease, resection may have a high risk of local recurrence and has a questionable response to chemotherapy. Role of local radiation therapy to decrease recurrence is not known yet.

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