ejpmr, 2017,4(4), 484-488

SJIF Impact Factor 4.161

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

EVAN'S TUMOR: A RARE CASE REPORT AND REVIEW OF LITERATURE

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Article Received on 10/02/2017 **Article Revised on 02/03/2017** **Article Accepted on 22/03/2017**

ABSTRACT

Evan's tumor or Low grade fibromyxoid sarcoma is a variant of fibrosarcoma with high potential for metastasis and sometimes a long interval between the tumor presentation and metastasis. We present the case of a 20-year-old male who developed a large mass in the sacral region. The tumor was excised. The tumor on histopathology evaluation was diagnosed as Evan's tumor. The disease recurred after a follow up of six months. The tumor was excised again and adjuvant radiation therapy was given to the patient. Due to the rarity of Evan's tumor, there is no dedicated follow-up protocol. In order to diagnose the disease and metastasis as early as possible, it is vital to inform the patients about the longstanding metastatic potential of the disease. Our patient, however, showed complete response to the treatment and is free of disease at six month follow-up.

KEYWORDS: Evan's, fibrosarcoma, metastasis.

BACKGROUND

Evan's tumor or low grade fibromyxoid sarcoma is a distinctive variant of fibrosarcoma. This pathologic entity was first described by Evans, and according to him, Evan's tumor is a rare soft tissue tumor with benign histologic appearance and high metastasizing potential.[1] Sometimes, long interval between the patient presentation and distant metastasis poses problems for the pathologists, radiologists, radiation oncologists and surgeons, with longstanding regular follow-up clinically and radiologically being the fundamental principle for tumor management. Although it is a well-described entity, still its exact incidence is difficult to estimate because of the paucity of the cases and diagnostic challenges. Usually, these tumors occur in the proximal extremities and trunk.[2] Less commonly, they may be found at rare locations, such as chest wall, retroperitoneum, or head.[2,3] The majority of Evan's tumors occur in subfascial location, but in rare cases, dermis or subcutis may be involved.[4] Evan's tumor typically involves young or middle-aged persons, but pediatric cases have also been described in the literature.[3,5-9] We present the case of a 20-year-old male who developed a large Evan's tumor at the sacral region. Imaging depiction, surgical approach of the mass and current follow-up recommendations is discussed.

CASE SUMMARY

A 20 year old male presented with low back pain and straining while defecation. MRI abdomen and pelvis revealed sacral mass. He underwent surgery for sacral spine tumor in April, 2016 and histopathological examination revealed haemangiopericytoma. The patient remained asymptomatic for six months. After that, he again presented with low backache, swelling, straining while micturition and defecation and numbness of perianal region; with no history of bowel or bladder incontinence. MRI lumbosacral spine with contrast revealed a large, heterogenous, multilobulated, soft tissue mass, centered on sacrum breaching right sacro-iliac joint. The mass was seen to cause severe narrowing of sacral spinal canal and mass extended into multiple sacral foramina. Posteriorly, non-enhancing / cystic extension from the mass was seen to extend into muscular plane with well defined margins (Figure 1, Figure 2). Anteriorly, the mass extended into presacral space. The mass was also extended into right piriformis muscle (Figure 3). Contrast enhanced computed tomography scan of neck, chest and abdomen revealed soft tissue mass based on sacrum causing destruction of bone with involvement of sacral canal and the mass showed extension into posterior sacral region with small presacral component with right piriformis muscle invasion. No significant lymphadenopathy was seen in rest of the body. He underwent re-exploration of lumbo-sacral region and decompression of recurrent large sacral tumor. Per operatively, the tumor appeared like a large, lobulated, yellowish, non suckable, avascular tumor in the pre- and paravertebral region extending into the sacral canal eroding the sacrum (S2). On gross examination, the cut surface was homogenous and pearly white with firm consistency. Microscopic examination revealed a low grade malignant mesenchymal neoplasm with well circumscribed pushing borders, covered with

thick fibrous pseudocapsules at places. The tumor showed variable cellularity, with moderately cellular areas showing haphazardly arranged and dyscohesive patternless sheets of fusiform to elongated tumor cells, in a background of loose fibromyxoid matrix showing a rich network of small ovoid thin walled blood vessels. The hypocellular areas revealed widely scattered similar tumor cells, within an edematous, hyalinised stroma showing slit like thin walled blood vessels. The tumor cells revealed stretched out eosinophilic cytoplasm with indistinct borders, few normal mitoses (1-2 per 10 HPF) and mildly pleomorphic ovoid to elongated nuclei with pointed to blunt ends, fine vesicular chromatin, smooth borders, and inconspicuous nucleoli. Foci of necrosis were not seen. Atypical mitoses or significant nuclear atypia were not seen. HPE was suggestive of Evan's Tumor (Low Grade Fibromyxoid Sarcoma). On immunohistochemistry, the tumor cells were found to be strongly positive for vimentin, but showed no expression for SMA, S-100, desmin and CD34. The MIB-1 labelling index within the tumor cells was 5-10%. He was then referred for post operative adjuvant treatment as Radiation therapy. After ensuring his routine blood investigations to be normal, he was taken for an aquaplast immobilization mask and a planning CT scan was done. Using CT simulation and Eclipse 3D treatment planning system, he was planned for Radiotherapy. He received 60 Gy in 30 fractions with 2 Gy per fraction to tumor bed, using 6 MV photons on linear accelerator. He received the treatment without much untoward complications. His routine blood counts were found to be normal throughout the treatment. After a follow up of six months, PET-CT whole body did not reveal any abnormal uptake. The patient remained asymptomatic.

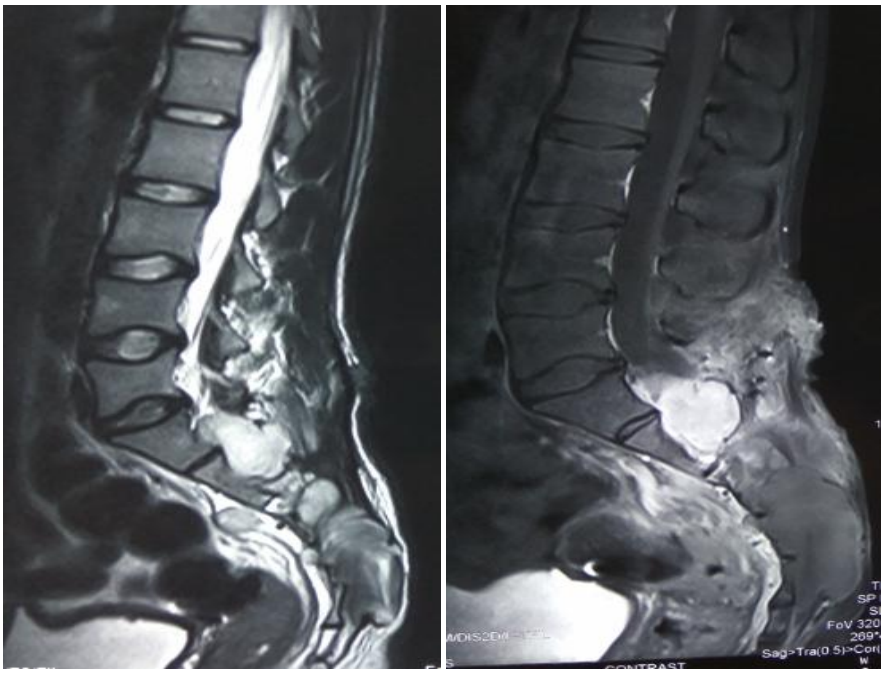

Figure 1. Sagittal MRI (T1W and T2W with ontrast) showing sacral mass with posterior extension of Evan's tumor

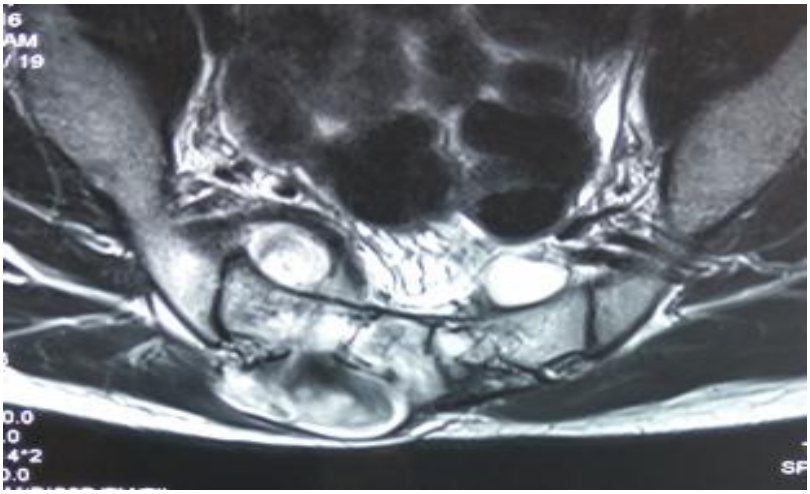

Figure 2. Axial MRI showing sacral mass with extension posteriorly

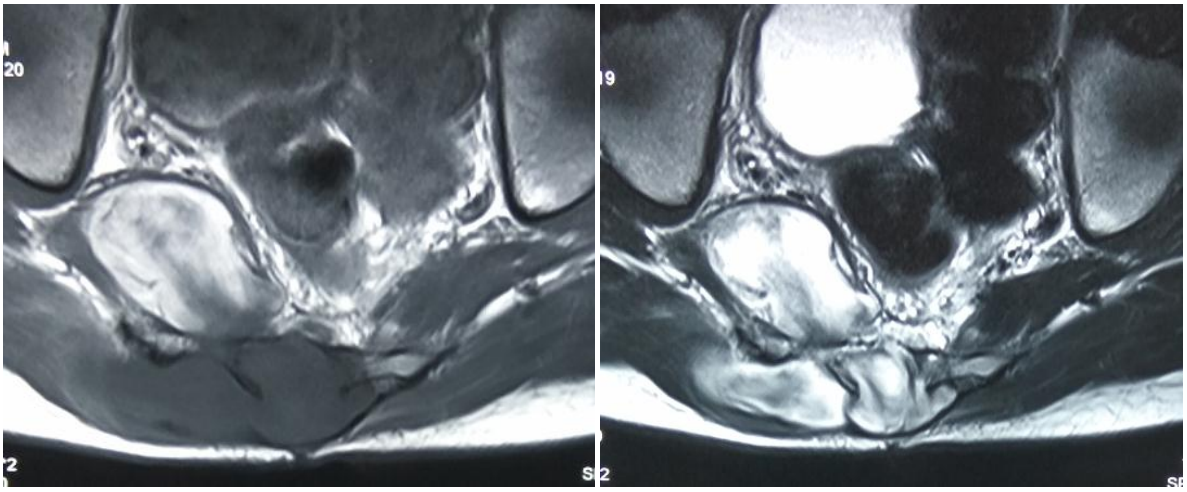

Figure 3. Axial MRI (T1W and T2W image with contrast) showing sacral mass with anterior extension to presacral space and right piriformis muscle involvement

DISCUSSION

Evan's tumor or low grade fibromyxoid sarcoma (LGFMS) is a characteristic variant of fibrosarcoma.[9] It is typically a cytologically bland malignant neoplasm with mixture of hypercellular myxoid nodules tangling with collagenized hypocellular zones.[2,10] This uncommon entity was described first by Evans.[1,6] Since then many cases have been reported of this rare tumor in the pediatric population.[11] The tumors vary in size from 1 to 23 cm with median size of 9.4 cm.[9,12] These tumors are usually located in the trunk and lower extremities, with shoulder region, thigh and inguinal region being the most common sites; and mesentery, thyroid and intracranial locations being the rare sites.[13,14,15] The Aarhus Sarcoma Registry found a 64% greater propensity among females.[16]

Tumor cells are peculiarly small, with meagre eosinophilic cytoplasm, nuclei is round to ovoid shaped and no nucleoli. Although tumor cells are characterized by sparse to absent nuclear anaplasia, mitotic figures or necrosis; still, cytologically focal atypical areas of increased mitotic activity, high cellularity, nuclear hyperchromatism and necrosis may be found in approximately 10% of the cases.[2] Immunohistochemistry is positive for vimentin and MUC4 only and negative for epithelial membrane antigen, keratin, desmin, S100 protein, CD34 and CD31.[1,17,18] Muscle specific actin is found to be positive in the walls of the small vessels within the tumor and strongly positive in the peripheral fibrous layer. A close pathologic entity, hyalinizing spindle cell tumor with giant rosettes (HSTGR) was reported ten years after the first description of Evan's tumor.[1,18] HSTGR tumors are characterized by proliferation of bland spindle cells with fibromyxoid areas. Giant rosettes are a distinctive pattern found in HSTGR and is defined by the presence of hyalinized acellular islands surrounded by spindle and oval cells. Folpe et al, in a large series of 77 cases of Evan's tumor and HSTGR, demonstrated that these tumors are part of the spectrum of low-grade sarcomas and represent the same neoplastic process.[9] They also revealed that several cases of Evan's tumor presented the pattern of miniature rosettes that had been overlooked at the time of the initial diagnoses. On the other hand, HSTGR is typically characterized by large areas that are histologically same to Evan's tumor. Thus, Folpe et al recommended that both entities should be referred to as "fibrosarcoma, low-grade fibromyxoid type" with a prescription about the rosettes, whether present or absent. The balanced translocation t(7;16)(q34;p11) and a fusion between the CREB3L2 and FUS genes is found in both Evan's tumor and HSTGR.[19-24] This translocation is peculiar for the diagnosis of these tumors and is typically useful in cases where discrete histopathological features are absent or in cases of restricted material for the examination. The FUS/CREB3L1 fusion transcripts of Evan's tumor can be reliably detected in paraffin embedded tissues using RT-PCR.[25]

Differential diagnosis of Evan's tumor includes lesions showing spindle cell proliferations with myxoid pattern with or without fibrous component.[4] The entities with primarily myxoid pattern without notable fibrous component include low-grade myxofibrosarcoma, myxoid liposarcoma, myxomas, angiomyxomas and myxoid neurofibroma. Tumors with mixed myxoid and fibrous morphologies include fibromatosis, neurofibroma, perineurioma, fibrous histiocytoma and malignant peripheral sheath tumor. Additional entities that should be encountered are low grade dedifferentiated liposarcoma, desmoid tumor and desmoplastic fibrosarcoma. The diagnosis of Evan's tumor or HSTGR is usually not difficult if the tumor has been removed completely and all the characteristic morphological and immunophenotypical features described above are present. This is not usually feasible when the material derives from fine needle aspiration or needle core biopsy.[4] In such cases, a wider biopsy, or even an excisional biopsy should be performed. If a myxoid pattern is there and still diagnosis remains uncertain,

cytogenetics must be performed to exclude Evan's tumor.[4]

The clinical presentation is usually related to the anatomic location of the mass and is mainly long-standing. Evan's tumor typically presents as a soft-tissue mass with no associated pain and over five years pre-biopsy duration in 15% of patients.[2] Acute appearance of the disease may occur in rare situations, such as chest pain and acute respiratory distress in Evan's tumor of chest wall or seizure episodes in intracranial Evan's tumor.[3,26] Whilst imaging findings are nonspecific for Evan's tumor, some undeniable CT and MRI findings have also been described.[3,26-28] On non-contrast CT images, fibrous component of Evan's tumor has been stated as isodense to muscle tissue and the myxoid element as hypodense. On MR imaging, the fibrous element is typically hypointense on T1- and T2-weighted MR images and mildly enhancing on T1-weighted MR images after gadolinium administration. On the other hand, the myxoid element is hypointense on T1-weighted MR images and hyperintense on T2-weighted MR images and vividly enhancing on T1-weighted MR images after gadolinium administration. Calcifications may also be found within the tumor.[26]

The mainstay of the treatment for Evan's tumor is surgery and techniques include local excision, wide en bloc resection, radical surgery and compartmental resection.[29,30] Margins are called as marginal if the resection included the pseudocapsule or wide if the resection included a cuff of normal tissue.[29] Local recurrences were treated with surgery without adjuvant therapy. However, distant metastases were treated with chemotherapy agents including trabectedin, doxorubicin, ifosfamide, gemcitabine, imatinib, docetaxel and radiotherapy. Trabectedin demonstrated the best response.[17] Evans and Goodlad et al suggested that Evan's tumor is paradoxically aggressive tumor.[6,8] In a retrospective analysis, local recurrence was seen in 68%, metastasis in 41%, and death in 18%.[2] Almost all these patients in these studies were initially diagnosed with and treated for a benign lesion. Metastasis was seen mainly to the lung followed by other soft tissues and very rarely in bone.[30,31] It is obvious that the patient selection has influenced the reported rate of recurrence and metastases, as many of those cases were selected on the base of unexplained metastases. In a recent series local recurrence, metastasis and death due to disease was observed in 54%, 6% and 2% of the patients, respectively.[9] This series also showed that the presence of focal areas of high cellularity, increased mitotic activity, nuclear enlargement and necrosis are not of prognostic significance for recurrent disease or metastasis. Given the potential of Evan's tumor for late metastasis, sometimes 45 years after initial diagnosis, the median follow up of only 24 months in the previous study should be considered too short in order to ascertain safe conclusions.[4] Once the diagnosis of Evan's tumor or HSTGR is made, a full oncological assessment should follow. This should include a CT scan of the chest, because pulmonary metastasis is the most obvious scenario. Because of the high risk of late metastasis, clinical follow-up and chest imaging must be done for a long period of time. However, it is still unclear how regularly imaging of the chest should be repeated.[4]

CONCLUSIONS

The case report presented herein, enriches the literature with information on imaging, diagnosis, surgical treatment and radiation therapy of this rare tumor. As there is no dedicated protocol regarding follow-up examinations and in order to early diagnose possible metastasis it is important to inform the patients about the longstanding metastatic potential of the disease. Furthermore, radiation therapy must be considered strongly in adjuvant setting post operatively to improve local control and to decrease distant recurrence or metastasis.

COMPETING INTERESTS

There are no competing interests.

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