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BMH Med. J. 2022;9(2):44-47. **Case Report**

Malignant Triton Tumor of Orbit - A Case Report

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Abstract

Malignant triton tumors are rare but highly aggressive tumors.They originate from the Schwann cells. These tumors can occur in any part of the body, they usually present in advanced stage and have very poor prognosis.

Keywords: Triton tumor, malignant peripheral nerve sheath tumors

Background

Malignant triton tumors (MTTs) are very rare subtypes of malignant peripheral nerve sheath tumors (MPNST) [1]. The reported incidence of sporadic MPNST and MTT is only 0.001% [2, 3]. MTT account for 5% of all MPNSTs [4]. They predominantly originate in the head, neck and extremities [2]. It can arise from different locations.

The prognosis of MTT is poor as reported by a 5-year survival rate of only 5 to 15% [5]. Literature has shown that the prognosis of MTT is influenced by many factors like its location (head, neck and extremities tumors carry better prognosis than the buttocks, retroperitoneal and central nervous system tumors), tumor size, degree of differentiation, tumor free resection margins, and Ki67 labeling index [6].

Case presentation

Our patient is a middle age gentle man with history of cerebrovascular accident at the age of 45 years, presenting with mass in left orbit with dimness of vision for 3 month. Surgical resection was attempted. Histology revealed inflammatory myofibroblastoma with malignant transformation. Pathology report came as spindle cell neoplasm. Within 3 months after surgery, patient again presented with orbital swelling. Debulking surgery attempted. The histological specimen composed of spindle cells, ovoid cells with lympho plasmocytic inflammatory component. Patient came with bleeding and crusting from the surgical site with in 3 months after debulking surgery. Orbital exentration of the tumor was done. Histology came as malignant triton tumor. Vimentin was diffusely positive. S100 showed diffuse positivity. SMA was

showing diffusely scattered positivity. Desmin was focally positive. After surgery patient defaulted and again came with mass in the orbital cavity. CT Head and Neck showed heterogenously enhacing mass causing erosion of medial wall and roof of left orbit, as well as it is extending into preseptal space. CT thorax, lung window was showing well circumscribed peripheral lung nodules suggestive of metastasis. Palliative radiation was given to locoregional site to control the disease. During radiation, patient developed middle cerebellar artery stroke and succumbed to death.

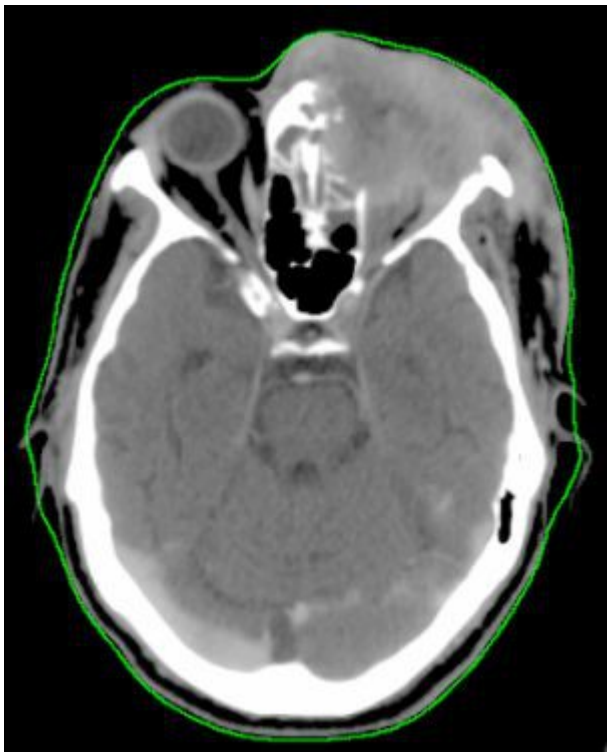

Figure 1: CT image showing heterogenously enhacing mass causing erosion of medial wall and roof of left orbit, as well as it is extending into preseptal space.

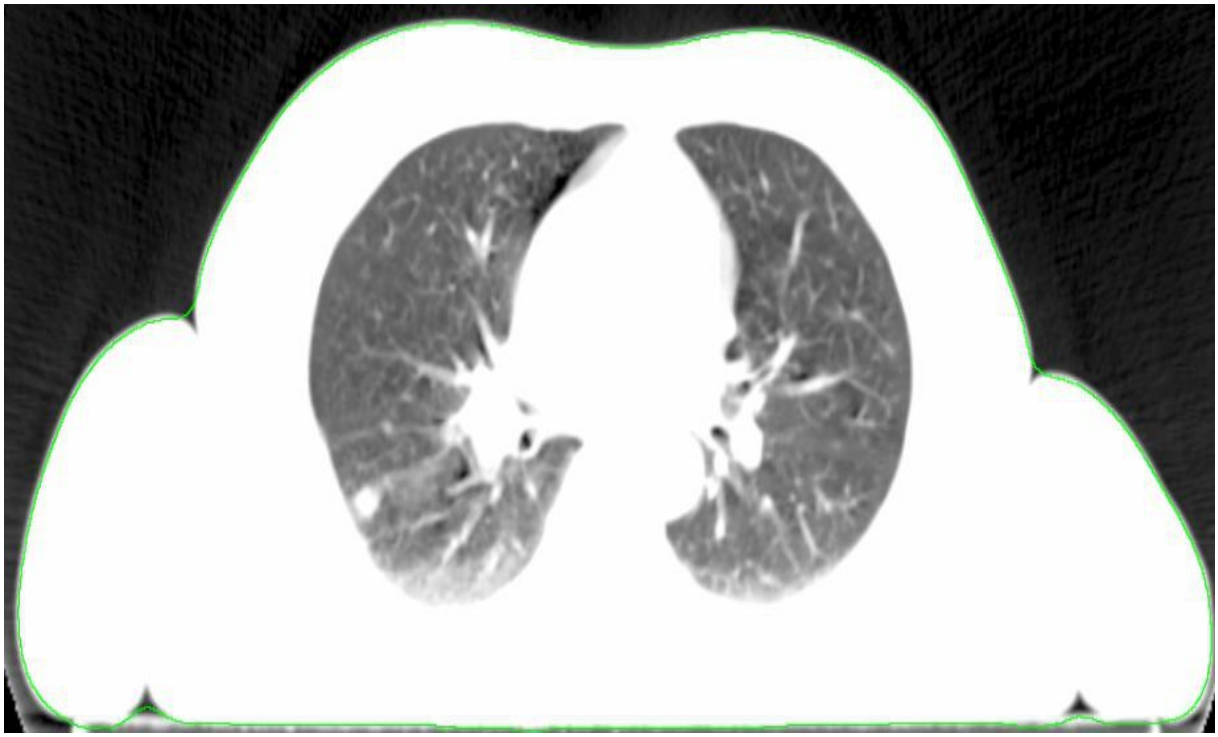

Figure 2: CT thorax, lung window image showing well circumscribed peripheral lung nodules suggestive of metastasis

Discussion

Very few (less than 200) cases of MTT have been reported in the literature worldwide [6,8]. MTT is a very aggressive tumor with its rate of distant metastases to be as high as 48% and a local recurrence rate of 43% [2,17]. MTT equally affects both men and women [5]. The mean age of presentation is 31.7 years [10]. The familial predilection in MTT is not reported and the same holds true for our patient as his past history was not remarkable of any familial correlation with this tumor. According to Woodruff et al., the diagnosis of MTT should meet three criteria [9]; first, the tumor originates from a peripheral nerve, in ganglioneuroma or in a patient with neurofibromatosis type I, or in a location typical for peripheral nerve tumors; second, the tumor demonstrates growth characteristics of Schwann cells; third, rhabdomyoblasts arising from the body of the peripheral nerve tumor. In addition to the criteria proposed by Woodruff et al., currently immunostaining is used for histopathological diagnosis of MTT. 50% of all MPNSTs staining for S-100 is positive. The positivity, seen in our case, is typically focal [11,12]. To make a diagnosis of a MTT, the presence of rhabdomyoblasts within an otherwise ordinary MPNST is very crucial.

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