Sclerosing Rhabdomyosarcoma Presenting in Masseter Muscle of an Adult: A Case Report

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Abstract— Sclerosing rhabdomyosarcoma is extremely rare skeletal muscle tumor which was recently included as a new subtype in 2013 by WHO of classification of tumors of soft tissue and bone.
Sclerosing rhabdomyosarcoma (SRMS) can be misdiagnosed and confused with osteosarcoma and chondrosarcoma by its histopathological pattern: lobules and infiltrating cords of small round malignant cells which embedded in a densely hyalinized matrix with osteoid and chondroid like appearance.
We report a case of SRMS presenting in masseter muscle of 29 years old Saudi male patient.
Although the imaging study of this patient suggested benign lesion with manifestation aswell circumscribed mass of right masseter muscle, the cytology suggested and histopathology confirmed the diagnosis as SRMS.

Index Terms— Rhabdomyosarcoma (RMS), sclerosing rhabdomyosarcoma (SRMS), pathology, immunohistochemistry, prognosis,computed tomography (CT), Magnetic resonance imaging (MRI).

INTRODUCTION

Rhabdomyosarcoma is a malignant soft tissue tumor that arises from a skeletal(striated) muscle cell. It is the most common single variety accounting for approximately 4.5% of all childhood malignancy with annual incidence of 5.3 per million children under the age of 15 years [3]...however, RMS can occur at any age being slightly more often in males than in females, with a ratio of approximately 1.3–1.5.

Rhabdomyosarcoma can occur anywhere in the body: Head and neck: about 40% of all rhabdomyosarcoma cases in children. This includes parameningeal sites, 25%; eye socket, 8%; and other head and neck locations, 7%. Urinary or reproductive organs: about 30% of cases.Arms or legs: about 15% of cases. Other sites: about 15% of cases. This includes the trunk (torso), intrathoracic, biliary tract, retroperitoneal, pelvic, and perineal sites.

(RMS) can be generally divided into three histological subsets:

Embryonal rhabdomyosarcoma which is the most common histological variant accounts (60-70%) of childhood cases, then are alveolar (20%) and pleomorphic (20%) variants.

The sclerosing/ spindle cell type was recently recognized by WHO of classification of tumors of soft tissue and bone in 2013. This type can be misdiagnosed and confused with osteosarcoma and chondrosarcoma by its morphological pattern: lobules and infiltrating cords of small round malignant cells which embedded in a densely hyalinized matrix with osteoid and chondroid like appearance.

The prognostic factors in RMS have been shown to be dependent on age, tumor site, resectability of the tumor, tumor size, regional lymph node involvement, the presence of metastasis, site, and extent of metastasis, and biological and histopathological characteristics of the tumor cells[4].

Case presentation :

A 29 years old Saudi male presented with right cheek swelling of three months duration, slowly growing, painless. Physical examination revealed a right masseter muscle mass, 3x3 cm firm in consistency , well defined and strictly attached to the muscle, no neck lymphadenopathy, no history of trauma or changes of overlying skin and no facial nerve injury.

MRI was done which showed, right masseter muscle demonstrating a well defined focal lesion of low signal intensity on T1W, inhomogeneous mixed signal on T2W, the measuring of lesion was (2.4 X 2 X 1.6) cm

CT was done showing small bilateral neck lymph nodes, the largest lymph node is noted at level II-A on the left side and measures 11mm on maximum short axis, swelling of right masseter muscle with a central area of low attenuation.

FNAC was attempted and reported as atypical cells.

Intraoperatively the tumor was unencapsulated and infiltrating with a firm to a hard consistency. On histopathology, sections showed infiltrating neoplastic lesion with tumor cells present in form of groups and infiltrating cords into the surrounding muscle tissue. The cells were medium in size and show vesicular nuclei with occasional prominent nucleoli. Few

scattered mitoses were also seen. Focal eosinophilic eccentric cytoplasm was seen. The surrounding tissue showed dense sclerotic areas.

Immunohistochemical stains applied with appropriate controls and tumor cells showed strongly positive for desmin, Vimentin, Myo-d1, MSA, while few cells were positive in Myogenin, Myoglobin, CD-34,

Ki-67 was expressed in 60% tumor cells. The tumor cells were negative for CD-31, CK AE1/AE3, EMA, LCA.

Morphological and immuno-histo-chemical features were consistent with the rhabdomyosarcoma-sclerosing variant.

Discussion :

Sclerosing rhabdomyosarcoma is an unusual variant of RMS especially in adult type, it was first described in 2000 by Mentzel and Katenkamp[2]. However so far, still there are few reported cases and information about the disease.

In our patient's case, he presented with a painless swelling in masseter muscular area with mildly enlarged lymph nodes and no intracranial invasion. It has been reported that the conventional subtypes of RMS in parameningeal locations could spread to the skull base or intracranial area, and this finding was related to poor prognosis [6]. Generally, SRMS has a good outcome in pediatric age group than in adults which has more risk for metastasis and recurrences.

The radiological investigation revealed mostly benign features of the tumor while FNAC and immunohistochemistry was consistent with Rhabdomyosarcoma – sclerosing variant

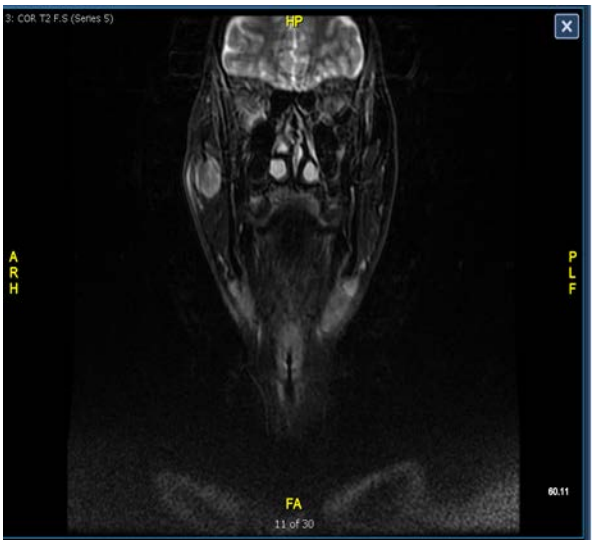


Fig:1 MRI Craniocervical Joint with + without Contrast

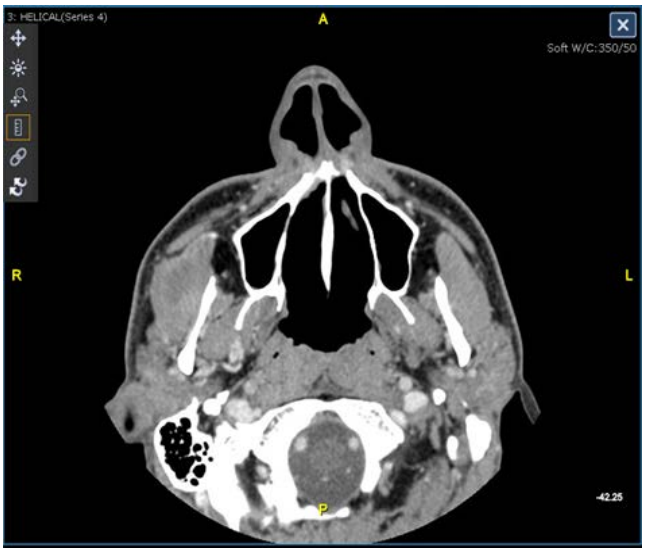
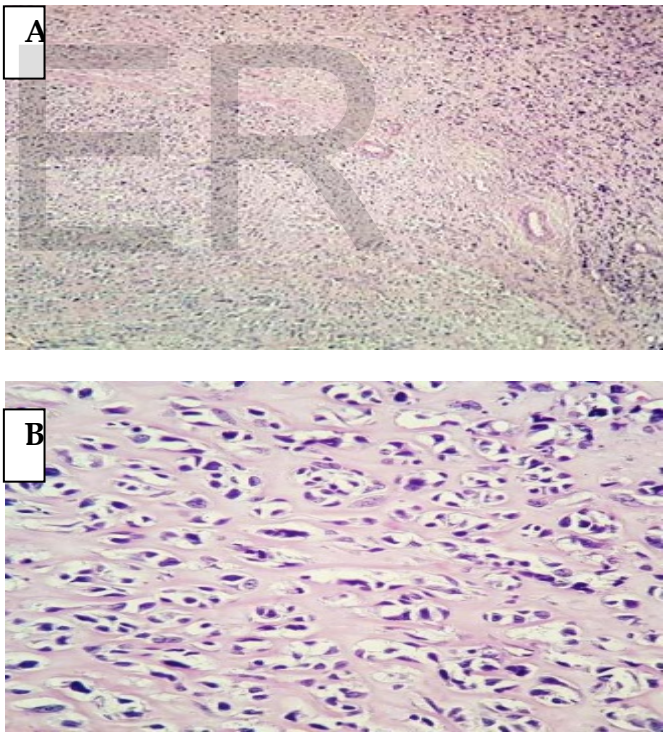


Fig:2 CT Maxillofacial with Contrast



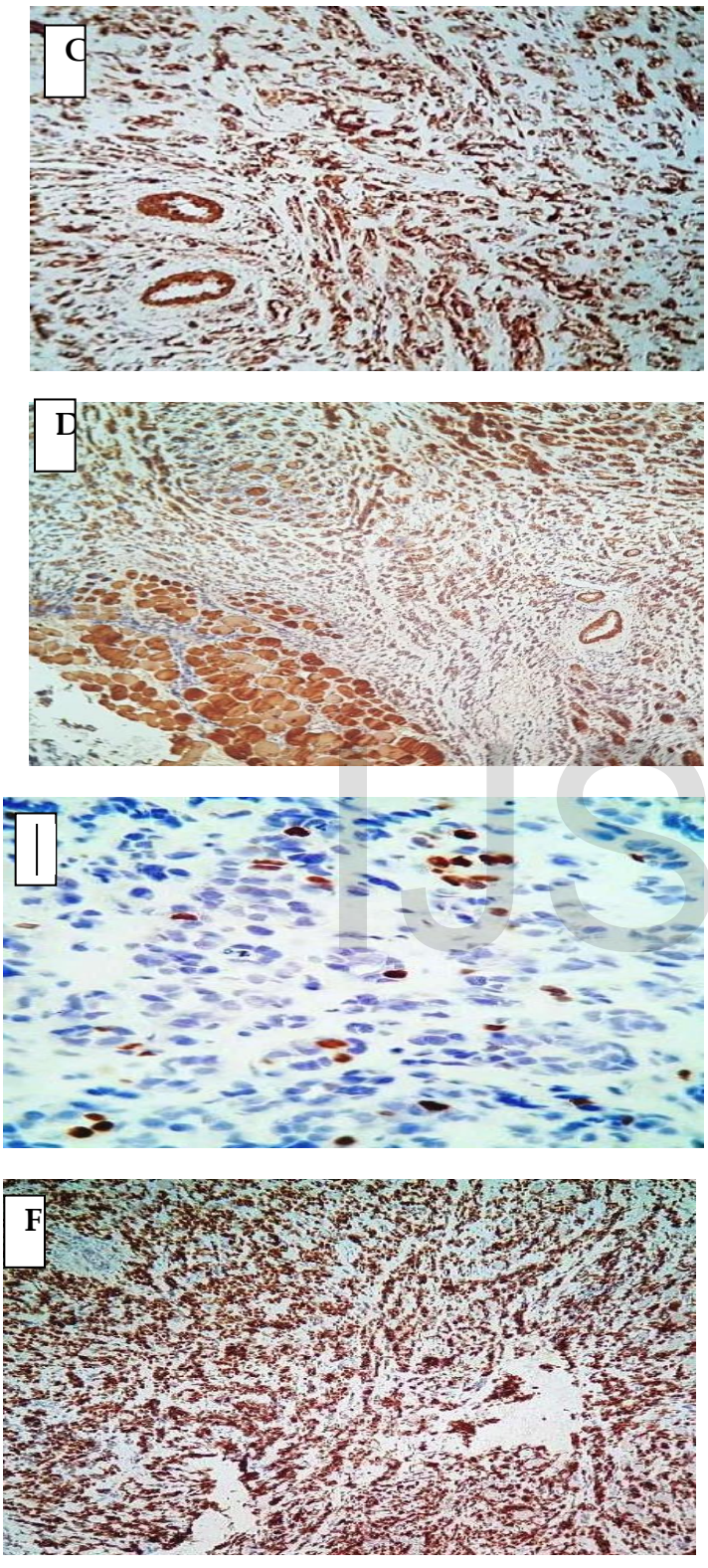


Fig.3: Hematoxylin and eosin (H&E) stain 20x, 40x (largest cells)the sections show (A) medium-sized tumor cells with vesicular nuclei and occasional prominent nucleoli, eosinophilic eccentric cytoplasm is seen. (B) the surrounding tissue shows dense sclerotic areas .on immunohistochemistry (C) MSA. (D) Desmin (E) Myogenin (F) Myo-D1 were positive.

The histological features of this case could be misdiagnosed due to overlapping features with other types of soft tissue tumor but the immunohistochemistry can help in differentiation. Definitive diagnosis also needs to rule out many differentials including osteosarcoma, leiomyosarcoma, fibrosarcoma, sclerosing epithelioid fibrosarcoma and other soft tissue tumors As in our case the tumor cells show strong and disuse positivity to Desmin, Vimentin, Myo-d1 while focal positivity to Myogenin, Myoglobin. Other RMS subtypes have different pathological features .

To avoid misdiagnosis, careful attention must be paid to its histopathological features.

Although the consensus of optimal treatment for SRMS has been not reached, the mainstay therapeutic method which is similar to most soft tissue tumors should be surgery and adjuvant chemotherapy or radiotherapy can be added [5]. the combination of surgery and adjuvant therapy greatly improve the prognosis.

Conclusion:

We report a rare case of Sclerosing RMS and because of the rarity, it can be misdiagnosed especially if one is unfamiliar with this entity. So, the pathologists need to keep this entity in mind in an adult population and do a proper workup, as the management varies from benign entities and other sarcomas.

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