Pleomorphic Rhabdomyosarcoma of Nose and Paranasal Sinuses in an Adult Patient: A Case Report

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Abstract

Pleomorphic rhabdomyosarcoma is a rare form of sarcoma in adults. It is commonly arises in the skeletal muscle of the extrimities with peak incidence in fifth decades with male predominance. It is uncommonly arises in head neck region which are aggressive tumour. Due to its variable histomorphology ancillary tests like immunhistochemistry is often necessary for diagnosis. We report a case of pleomorphic rhabdomyosarcoma of nose and paranasal sinuses in 34 years old female with lymph node metastasis. Histopathological examination showed sheets of tumour cells having round to spindly nuclei with small amount of cytoplasm and large atypical multinucleated polygonal cells. Diffuse cytoplasmic desmin positivity and nuclear myogenin positivity confirmed the diagnosis. After surgery patient took chemotherapy and died after six months.

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Key words: Pleomorphic rhabdomyosarcoma, immunohistochemistry, nose and paranasal sinuses, soft tissue tumour

Introduction

Pleomorphic rhabdomyosarcoma is a rare type of sarcoma in adult which gained popularity and reported during the period of 1930s and 1940s.[1] According to WHO classification of tumors of soft tissue tumor and bone rhabdomyosarcoma is subtypes as embryonal, alveolar, pleomorphic and spindle cell/sclerosing.[2] Among these subtypes embryonal rhabdomyosarcoma is the most common variant predominantly affecting children whereas pleomorphic rhabdomyosarcoma occurs in adults with a peak incidence in fifth decade of life with a predilection for males.[1] Commonly this tumor arises in the skeletal muscle of the extrimities particularly thigh. Less common sites are abdomen/retroperitoneum, chest/abdominal wall, spermatic cord/testis and upper extrimities. Rare cases may locate in the mouth and orbit.[3]

Ancillary techniques like immunohistochemical detection of sarcomeric differentiation using antibodies to desmin, muscle specific actin, myo D1 and myogenin is essential to categorize it from other sarcoma. In cytogenetics most of the pleomorphic rhabdomyosarcoma have a highly complex karyotype. However it does not have any characteristic aberration.[1]

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Case report

We are reporting a case of pleomorphic rhabdomyosarcoma involving both nasal cavities, right maxillary antrum and right ethmoid region. Patient was a 34 years old female, presented with the complaints of headache, right nasal blockage and swelling on the right side of the face over maxillary region (Figure 1) including eyeball for one and half months. With this problem patient was referred from a district hospital to Bangabandhu Sheikh Mujib Medical University (BSMMU). Initially she has underwent biochemical and radiological investigations. Her Hb% was found low, other biochemical tests were within normal limit.

CT scan of brain and orbit showed enhancing soft tissue density area originating from the ethmoid sinuses extending posteriorly into sphenoid sinuses. The mass also extends antero-inferiorly into right maxilla and both nasal cavities. Radiologists reported as right sided sinonasal mass with mild proptosis of right eye ball and normal brain. CT scan of paranasal sinuses showed large infiltrative mass of ethmoid sinuses associated with extensive bony destruction and right orbital with intracranial extension (Figure 2). Biopsy was taken from right nasal cavity. Grossly biopsy sample was consisting of 0.8 cc multiple irregular grey white pieces of tissue. Morphologically a malignant tumour was found which revealed cells having round to spindly nuclei with small amount of cytoplasm.

Differential diagnosis was olfactory neuroblastoma and undifferentiated carcinoma. With this patient underwent surgery. Per operative findings showed that the tumour had involved maxillary antrum, upper part of nasal cavity and right ethmoidal region. Right medial maxillectomy with right sided radical neck dissection was performed. The resected sample was received in Pathology department where tumour was found as multiple grey white irregular fragmented pieces of tissue measuring in between 2x2x0.3 cm to 0.5 cm with an irregular piece of 1.0 cm bone. Radical neck dissection sample was received where submandibular salivary gland and three lymph nodes measuring in between 2 cm to 0.5 cm was found. Nine lymph nodes were dissected from right cervical lymph nodes of level II, III and IV. In resected sample, histopathology of tumour revealed sheets of tumour cells having round to spindly nuclei with small amount of cytoplasm (Figure 3) and many large atypical cells often with multinucleated polygonal forms (Figure 4).

Immunohistochemistry for pancytokeratin, EMA, S-100 protein, neuron specific enolase, synaptophysin, HMB45, Melan-A, Leukocyte common antigen (CD45), desmin and myogenin was performed. Tumour cells were positive for desmin, myogenin (Figure 5, 6) and neuron specific enolase. Final diagnosis was made as pleomorphic rhabdomyosarcoma, grade III with metastasis in right submandibular and right cervical lymph node.

Postoperative period of patient was uneventful and was recovered quickly. Chemotherapy was given afterwards but patient was died after six months of her surgery.

Figure 1. Female, 45 years. Huge swelling on the right side of face

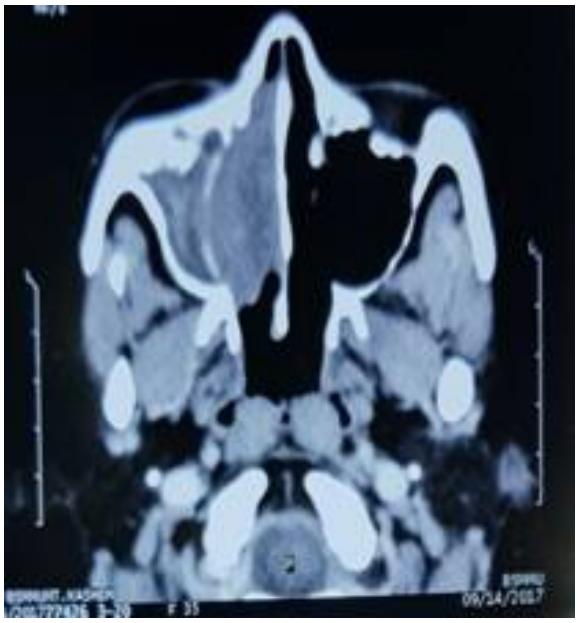

Figure 2. CT scan of paranasal sinuses shows large infiltrative mass of ethmoid sinuses

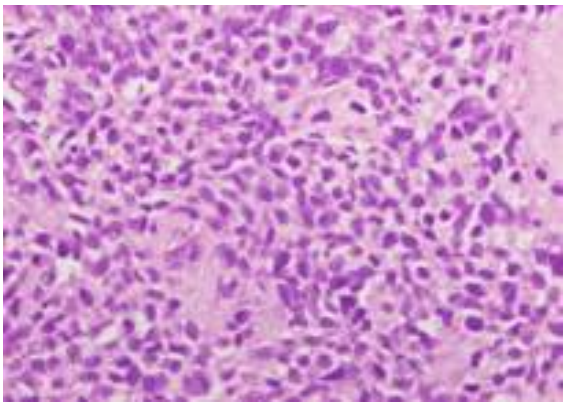

Figure 3. Histological section shows malignant cells having round to spindly nuclei with small amount of cytoplasm (H & E Stain, x400).

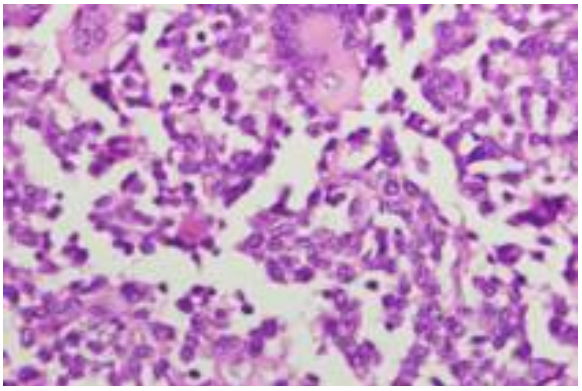

Figure 4. Histological section show many large pleomorphic cells often with multinucleated polygonal cytoplasm (H & E Stain, x400).

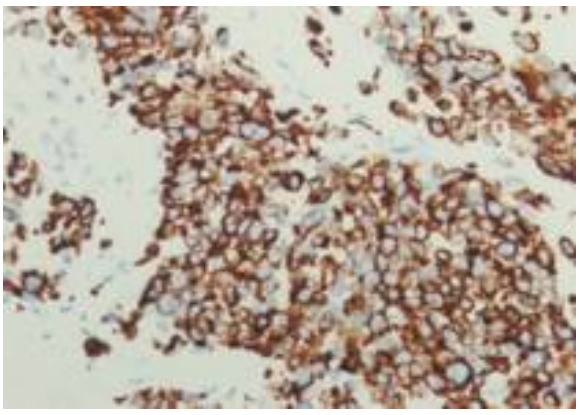

Figure 5. Immunohistological stain reveals positive reactivity of tumour cells to desmin (IHC Stain, x400).

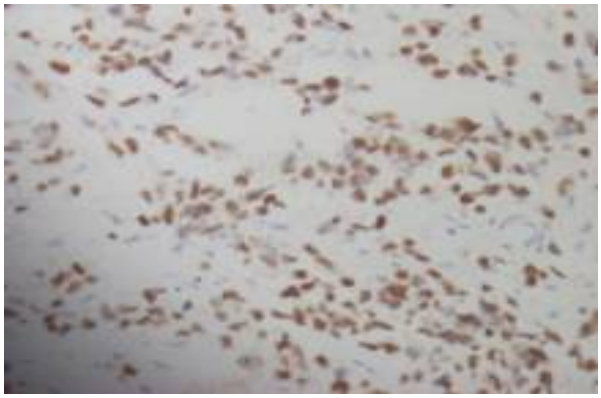

Figure 6. Immunohistological stain reveals positive reactivity of tumour cells to myogenin (IHC Stain, x400).

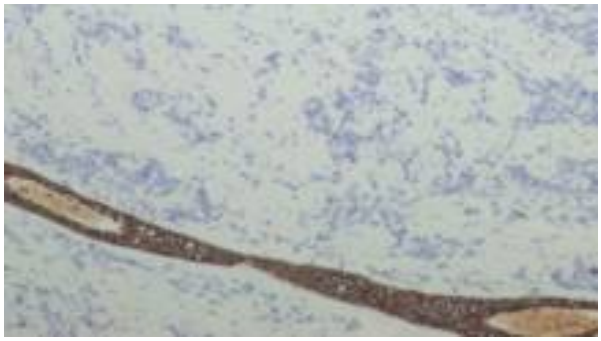

Figure 7. Immunohistological stain reveals negative reactivity of tumour cells to pancytokeratin (IHC Stain, x100).

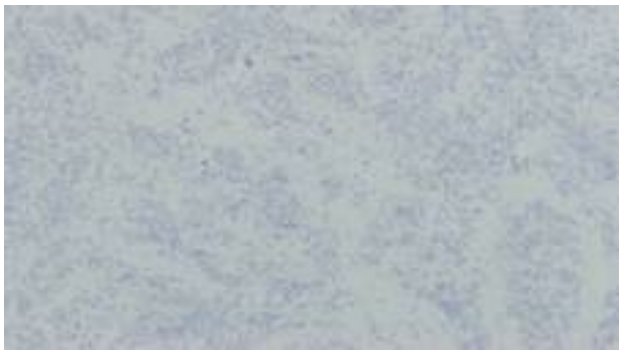

Figure 8. Immunohistological stain reveals negative reactivity of tumour cells to S100 protein (IHC Stain, x100).

Discussion

Among soft tissue sarcoma rhabdomyosarcoma is the most common in children representing 5% of all childhood cancers. In contrast, in adults it is rare, accounting for <1% of all malignancies and accounting for 3% of all soft tissue sarcomas.[4,5] Among the histological subtypes embryonal rhabdomyosarcoma accounts for 60% in children whereas 5% are pleomorphic rhabdomyosarcoma. Though rhabdomyosarcoma is relatively rare in adults pleomorphic rhabdomyosarcoma is most common subtype in comparison to other subtype.[3,6]

The most frequent site of involvement of pleomorphic rhabdomyosarcoma is the extrimites. On the other hand, least common site is head-neck region.[3] Anatomically, rhabdomyosarcoma are classified as parameningeal, orbital, nonparameningeal and nonorbital. The parameningeal sites include the nose, nasopharynx, paranasal sinuses, middle ear, mastoid, infratemporal fossa and pterygopalatine fossa and tumour of these sites carry the worse prognosis. We reported a case of pleomorphic rhabdomyosarcoma involving right maxillary antum, nasal cavity and right ethmoidal region of a 34 years old female having aggressive clinical behavior.

Regarding morphology of pleomorphic rhabdomyosarcoma Furlong et al[3] has described three morphological varients as classic pleomorphic rhabdomyosarcoma, round cell pleomorphic rhabdomyosarcoma and spindle cell pleomorphic rhabdomyosarcoma. Classic variant is described as tumor consisting of sheets of large, atypical and often multinucleated polygonal pleomorphic rhabdomyoblasts. Round cell revealed large atypical pleomorphic rhabdomyoblasts, throughout the lesion with a background of slightly pleomorphic medium-sized, round, blue rhabdomyoblasts and spindle cell varients consisting of predominance of pleomorphic spindled pleomorphic rhabdomyoblasts

arranged in a storiform growth pattern with scattered large pleomorphic rhabdomyoblasts.[3] Histological feature of our case has similarity with round cell type. This is the possible explanation of initial nasal biopsy to be reported as olfactory neuroblastoma with differential diagnosis of undifferentiated carcinoma.

Due to the location of tumour in the ethmoid sinus with extension in the maxilla, both nasal cavity and sphenoid sinus complete excision was not possible. In the resected sample areas containing pleomorphic cells are found with a background of medium sized round to spindle shaped cells. Diffuse cytoplasmic desmin positivity (Figure 5) and nuclear positivity of myogenin established these cells as rhabdomyoblasts (Figure 6).

Due to rarity of this tumour histomorphological features need to be supported by other ancillary tests. Our case showed desmin and myogenin immunostain positivity which is similar to the report of Furlong et al in their clinicopathologic study of pleomorphic rhabdomyosarcoma in adults.[3] Present case showed diffuse strong cytoplasmic positivity of desmin which was observed by Caporlingua et al, Crane et al, Mungan et al, Yu et al Deniz et al and in their reported cases of pleomorphic rhabdomyosarcoma in various location.[8-12] Nuclear positivity of myogenin was reported by Caporlingua et al, Crane et al, Mungan et al and Deniz et al. [8, 9,10,12]

As morphology of the present case showed round to spindly cells with scanty cytoplasm with areas of pleomorphic cells, differential diagnoses was included undifferentiated carcinoma, olfactory neuroblastoma, pleomorphic sarcoma, malignant melanoma and lymphoma. Immunohistochemistry for pancytokeratin (Figure 7), EMA, S-100 protein (Figure 8), synaptophysin, HMB45, Melan-A, Leukocyte common antigen(CD45) negativity in tumors has excluded the differential diagnoses of undifferentiated carcinoma, olfactory neuroblastoma, malignant melanoma and lymphoma.

Patients with rhabdomyosarcoma are treated with surgery followed by radiotherapy and chemotherapy. Surgery is the main modality of primary procedure or a secondary strategy after initial chemotherapy.[5] Several prognostic indicators like age, tumor location, and histotype have been identified for rhabdomyosarcoma.[9] There are reported cases of better prognosis for patients aged <20 years, with a tumor size <5 cm, treated with radical surgical excision in the absence of regional lymph node involvment.[13]

In our reported case patient's age, tumours location in the nose and paranasal sinuses, histologic type of pleomorphic rhabdomyosarcoma with nodal metastasis have carried aggressive clinical behavior.

Conclusion

Pleomorphic rhabdomyosarcoma is a rare type of sarcoma in the head neck region with variable morphology. Immunohistochemistry is an essential ancillary test for its accurate diagnosis. Appropriate panels of immunomarker selection are important for conclusive diagnosis.

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