CASE REPORT

An infrequent case of adult nasopharynx rhabdomyosarcoma with bony involvement: A case report

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Abstract

Rhabdomyosarcoma is a rare and highly aggressive malignancy arising from the striated skeletal muscle fibre. It is commonly a childhood tumour and rarely seen in adults. The most predominant part commonly involved is head and neck. We are reporting a rare case of a 54 year old adult male with Rhabdomyosarcoma of nasopharynx with a non specific presentation of weight loss for past four months, fever, a perianal abscess and backache since 10 days. The haematological and biochemical parameters were within normal limits however C reactive protein and erythrocyte sedimentation rate were raised. An MRI for the pelvis, which was done for the extension of perianal abscess, also displayed some heterogeneous marrow signal, which raised the suspicion of some infiltrative process. Later, a bone scan, PET scan, and a biopsy of the nasopharaynx was done. The biopsy confirmed the findings of Rhabdomyosarcoma by showing marked pleomorphism with strong positive desmin and myogin stain. The patient was then referred to the oncology department for further management.

Keywords: Rhabdomyosarcoma, nasopharynx, Backache,

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Introduction

Common malignant tumours of the musculoskeletal system in children, adolescents, and young adults (<30 years old) include rhabdomyosarcoma, osteosarcoma, and Ewing's sarcoma.[1] Together, they constitute about 10 percent of newly diagnosed cancers in the age group of people who are less than 30 years (about 1000 cases yearly in the United States)however, the peak age is 2-6 years. Although relatively common in young persons, these tumours are infrequent in elderly people.[2] RMS can occur in any region of the body, but is commonly found in the head, neck, orbit, genitourinary tract, genitals, and extremities. The presentation depends on the region involved.[3] There are no clear risk factors for Rhabdomyosarcoma. It has been derived from primitive undifferentiated mesenchymal cells and has been classified into embryonal, alveolar, and pleomorphic categories. Anatomically, RMS are classified as parameningeal, orbital, nonparamenigeal, and nonorbital. The parameninngeal tumour carries the worst prognosis.[4] Rhabdomyosarcoma is often difficult to diagnose, because of its resemblance with other malignancies of young age like neuroblastoma, Ewing sarcoma, and lymphoma. Biopsy is required for confirmation, with immunohistochemical stain specific for muscles that is Myogen, Desmin, D-myosin, and myo D1. Bone scan, PET scan, and MRI further guide us to know the extent of the disease.[5] The purpose of this article was to report a case of nasopharangeal rhabdomyosarcoma which is infrequent in adult population and to discuss the clinical, radiological, and histopathological challenges.

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Case Report

The case was first seen in the rheumatology clinic of DOW University of Health Sciences, Ojha Campus, Karachi in July 2019.The consent of the patient was taken prior to the writing of the manuscript. The patient was a 54 year old married male and a known case of diabetes for the past four years. He was referred from Nawabshah to our clinic with the complaint of fever and backache since 10 days. Fever was intermittent and was documented to be up to 103°F. He had a history of weight loss of 4 kg in 4 months and occasional rectal bleed, with last bleeding episode 10 days prior to admission, He had a history of single episode of epistaxis a month ago. He denied the history of altered bowel habits, cough, nasal obstruction, and rhinorrhoea. His contact history of tuberculosis was positive. On examination, tenderness was present at the thoracic region, deviated nasal septum was also noticed. Haematological and biochemical parameters were within normal limits, however, CRP and ESR were raised. (CRP 267mg/L,ESR 110 mm/hr).The reference range of CRP is 0-10mg/L and the reference range of ESR is 0-15mm/hr.Urine DR, Stool DR ,Chest X-ray, X-ray lumbo sacral spine, Ultrasound abdomen, and Serum electrophoresis, were all unremarkable. MRI cervical and lumbo sacral spine showed degenerative changes. A surgical consult was generated for peri anal abscess and MRI pelvis was advised, which showed subtle enhancing tract within the inter-sphincter region in the midline with internal mucosal opening at 6 o'clock position in the anal canal and extending in the midline in the natal cleft, however, no definite external

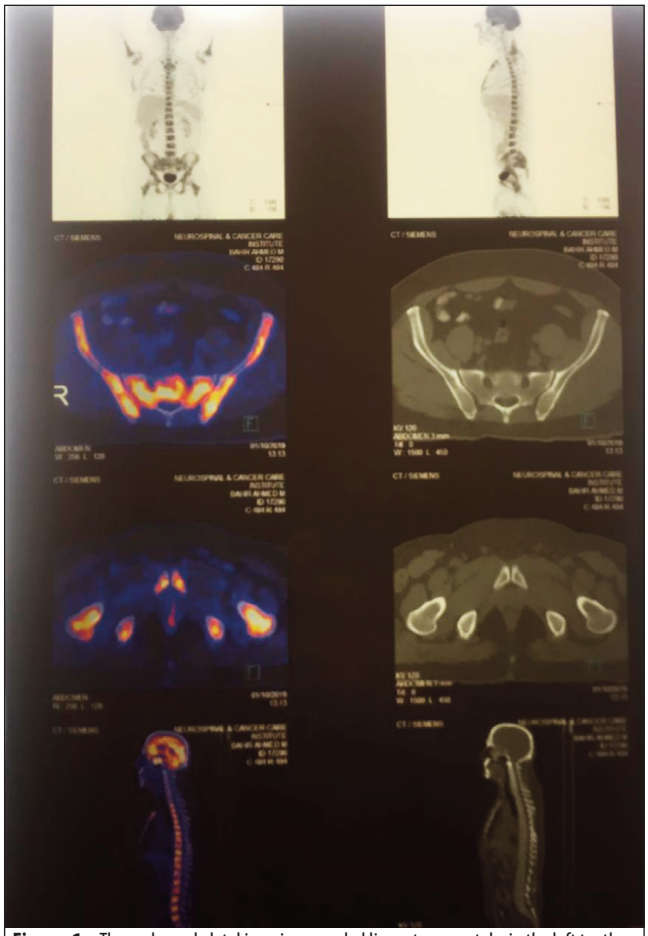

Figure-1: Three phase skeletal imaging revealed linear tracer uptake in the left tenth rib and inferior angle of the right scapula.

opening was seen. There was an associated mild reactive mucosal enhancement. The visualized pelvic bone predominantly acetabulum femur and the visualized spine showed heterogeneous marrow signals, revealing patchy post contrast enhancement. The prostate was of normal size and configuration. No evidence of mass lesion or abnormal signals were noted. A few prominent bilateral inguino pelvic lymph nodes were seen. Fistulectomy was performed during the hospital stay. Three phase skeletal imaging revealed linear tracer uptake in the left tenth rib and inferior angle of the right scapula. PET SCAN revealed evidence of FDG avid soft tissue thickening, seen in the sphenoid sinus. The lesion measured 3.1*2.9*2.4cm. (figure 1 and 2). The findings were suggestive of a metastatic disease of the bone with possible primary site being the nasopharynx or prostate. Prostate specific antigen was normal and patient was sent to ENT department for fibroptic direct laryngoscopy and biopsy. Immuno-histochemical stains were all in favour of Rhabdomyosarcoma showing Desmin strong positive, Myogenin strong positive, CD 117, and PLAP weak positive

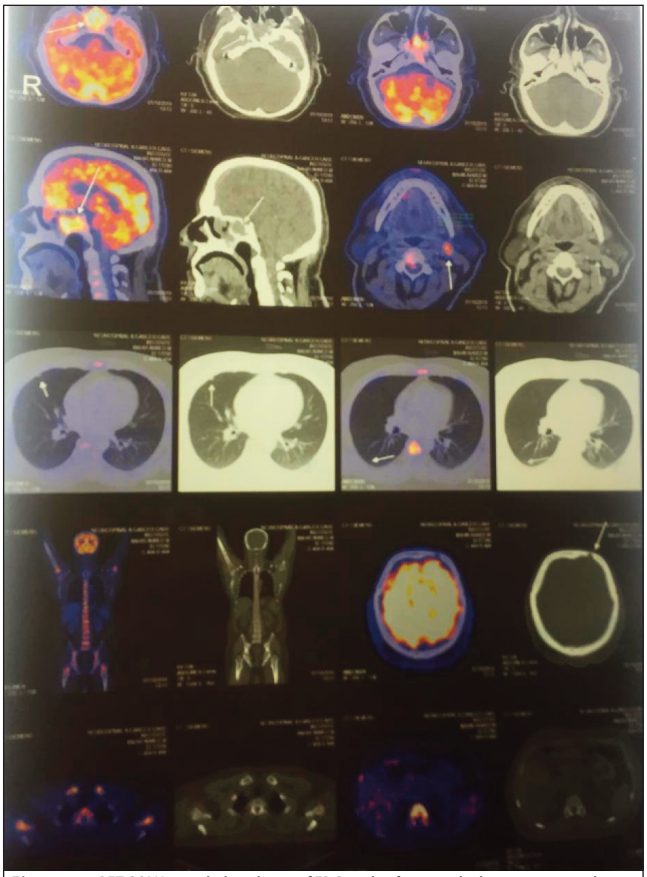

Figure-2: PET SCAN revealed evidence of FDG avid soft tissue thickening, seen in the sphenoid sinus.

.The patient was then referred to an Oncologist for further management, Later, the bone marrow was also examined which exhibited its extension .The patient is now on follow-up of an oncologist for regular check ups, after one cycle of VAC. Up till now the patient has not achieved remission.

Discussion

Rhabdomyosarcoma is a rare soft tissue malignancy of the striated muscle fibre and constitutes only 3% of all soft tissue sarcomas.[6] Our case reflects the importance of early MRI and PET scan when the lesion is suspected to be. Rhabdomyosarcoma. This tumour is a challenging diagnosis, often difficult to reach because of its non-specific symtoms and atypical presentation. The prevalence of bony metastasis in Rhabdomyosarcoma is not clearly defined. This malignancy has not specified salient imaging characterstics. Musculosketal invasion was displayed in 24% of cases. The bone metastases are typically ill-defined lytic lesions.[7] We do consider this case as bony metastasis, with the primary being in the nasopharynx, and extensive metastasis to the bone as well as marrow. Bone marrow lesions are found in about 30% of the metastatic cases at presentation. In our case, we also found bone marrow

metastasis. Bone marrow metastases at the time of diagnosis signifies worse prognosis than with the metastases to other vital structures.[8]

A similar case of rhabdomyosarcoma of nasopharynx was published in 2005 in a 67 year old male without any nasal symptoms. The patient achieved complete remission with combination chemotherapy.[9] A case report of an adult orbitalethmoidal rhabdomyosarcoma with intracranial extension published in 2014, did not achieve complete remission and was kept for palliative care.[10] A similar case was reported from Australia, highlighting the importance of MRI and identifying bony metastasis in a fifty year old male presenting with acute urinary retention. CT scan revealed protusion of L4 and L5. The patient expired after 1 week of diagnosis.[11] Metastasis of primary adult rhabdomyosarcoma of nasopharynx has also been detected in the breast, 17 months after complete remission.[12] No adult rhabdomyosarcoma of nasopharynx with metastasis has been reported locally so far. This case report is the first of its kind due to its unusual and infrequent presentation. A multidisiplinary treatment approach is required for the management of rhabdomyosarcoma, including surgery, and chemoradiotherapy. With all the combined effort, the survival rate of Rhandomyosarcoma can be improved.

Conclusion

Our case highlights the importance of radiological investigation in cases of diagnostic challenge. Physician should be aware of the various asymptomatic and atypical presentation of rare malignancies, so that timely biopsy and other measures can be taken.

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Conflict of Interest: None.

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