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CASE REPORT
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EXTRA SKELETAL EWING'S SARCOMA OF SINONASAL TRACT – A RARE ENTITY

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ABSTRACT

INTRODUCTION

Ewing's sarcoma of sino-nasal tract is an uncommon entity belonging to the Extra skeletal group of Ewing's Sarcoma (EFST) and is predominantly seen in young adults with mean age of 15-20 years(1).

CASE REPORT

We report a case of 16 years old boy who presented with complaints of right nasal obstruction, facial pain , swelling, mucosal discharge and episodes of epistaxis. On radiological work up possibility of juvenile Angiofibroma / neoplastic mass was suggested. On histopathology, diagnosis of Ewing's sarcoma was made. The treatment of choice is mainly surgery , chemotherapy and radiotherapy(2).

CONCLUSION

Ewing's Sarcoma originating from sinonasal tract is a rare entity .It is seen mainly in adolescent males. The main stay of diagnosis is histopathology coupled with immunohisto-chemistry and extent can be delineated by contrast enhanced CT neck.

CASE REPORT:

A 16 years old adolescent male presented with unilateral nasal obstruction since 3 months along with facial swelling, pain and right nasal mucosal discharge of 2 months duration. He also had multiple episodes of epistaxis in the last10 -12 days. Anterior rhinoscopy revealed red polypoidal soft tissue mass in the right nasal cavity with surrounding blood clots as shown in figure 1.

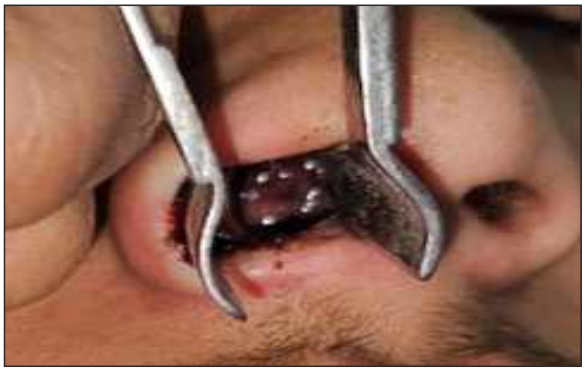

Figure 1

A class of segmented worms known as leeches comprises over 600 species, among which only a minority are sanguivores Leeches are frequently encountered in wilderness environments, typically by explorers and travelers, owing to their propensity for external

Contrast Enhanced CT(CECT) of Para Nasal Sinuses shows presence of a heterogeneously enhancing polypoidal soft tissue measuring 3.7x3.7x4.2cm(APXTRXCC) with average attenuation value of 40-60 HU seen in the right maxillary sinus extending into the right nasal cavity through the right OMC ,causing its expansion and obliteration as shown in figure 2.

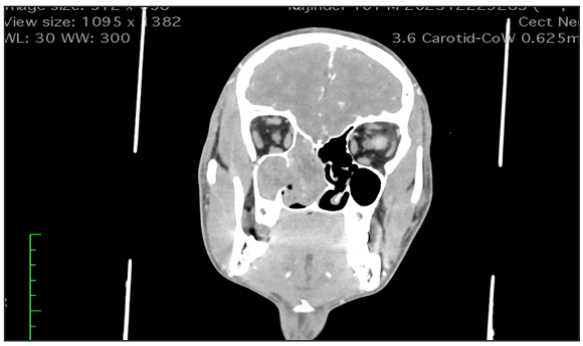

Figure : 2

Superiorly, it is extending into right loculus of the sphenoid sinus and extending intracranially by erosion of sphenoid bone in the basifrontal region anterior to the sella .It is causing thinning and expansion of the posterior lateral wall of the right maxillary sinus and superior, middle and inferior turbinates as shown in figure 3.

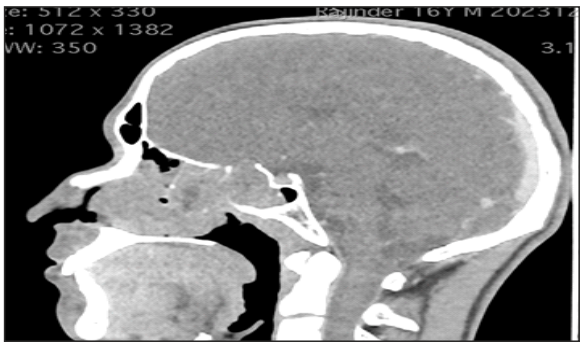

Figure: 3

Posteriorly, it is reaching upto the nasal choana causing its obstruction; however no extension into the nasopharynx is seen as shown in fig. 4.

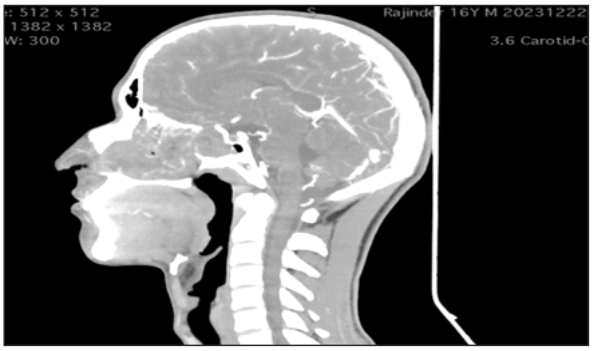

Figure: 4

It is seen extending into the right Sphenopalatine foramen, causing its widening and minimal extension into pterygopalatine foramen as shown in figure 5.

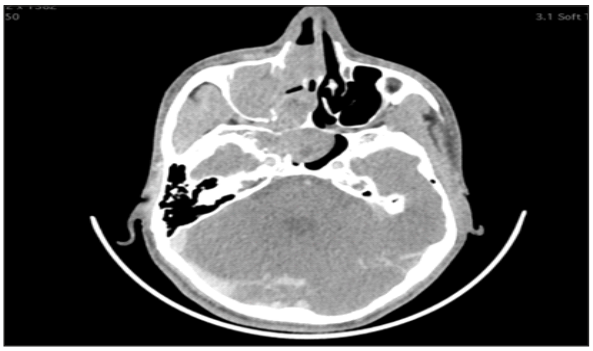

Figure : 5

A digital subtraction angiography was attempted to rule out juvenile Angiofibroma a potential diagnosis owing to recurrent epistaxis and involvement of Sphenopalatine foramen in an adolescent male.

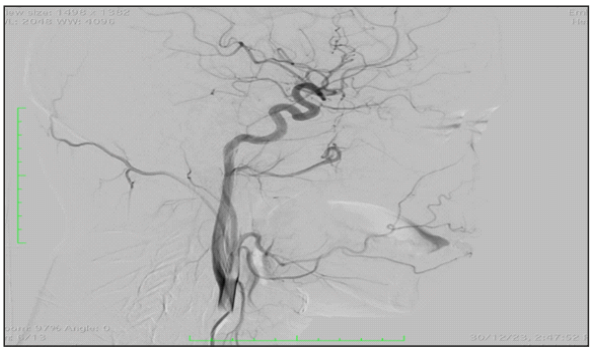

Figure : 6

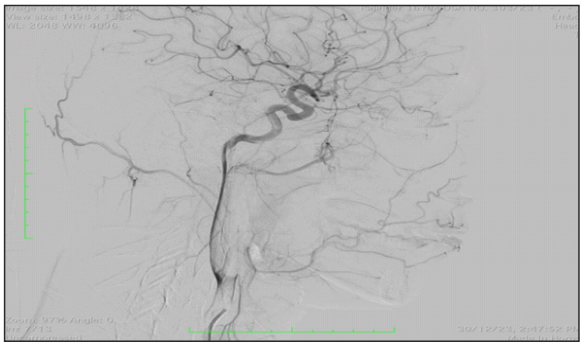

Figure : 7

Rapid injection of the iodinated contrast into the right External carotid artery and its branches did not show any significant blush of contrast in the sphenopalatine branches of the right maxillary artery as shown in figure 6 and 7 .

Thus possibility of juvenile angiofibroma was ruled out.

Histopatholgy of the mass was performed which revealed normal overlying epithelium beneath which were presentsheets, cords, pseudo-rosettes and islands of tumor cells separated by fibrous septa .The tumor cells showed high N/C ratio, small sized, round to irregular nuclei with stippled chromatin, inconspicuous nucleoli and scant to moderate eosinophilic granular vacuolated cytoplasm. On immunohistochemistry, tumor cells showed strong diffuse membranous immuno-reactivity for CD99 and Ki 67 index of 30-40 % consistent with Ewing's sarcoma of right nasal cavity as in figure (8 &9).

Desmin and CD45 were non-immuno reactive.

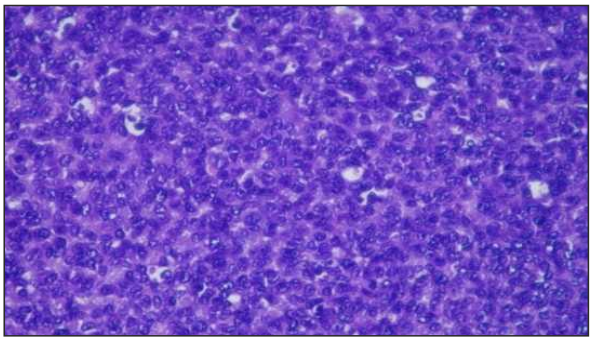

Fig 8 (Small blue round cell tumor with pseudorosettes, H&E stain, 200x)

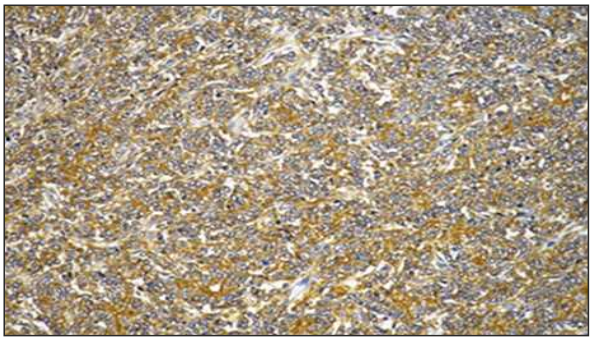

Fig 9 (Diffuse membranous positivity with CD99,100x)

Local excision and removal of the mass was done and the patient was discharged and doing well.

DISCUSSION

Ewing's sarcoma of bone was first described by James Ewing's in 1921(10). It is a small round tumor known to occur in less than 30 years of age with peak incidence between 10 to 15 years(11,12).

Ewing's sarcoma of sinonasal tract is a very rare entity in adolescent males .It is highly malignant in nature and known to infiltrate the surrounding sinuses and soft tissue with bone erosions in few cases(8,9).

Clinical manifestations of the Ewing's sarcoma of the sinonasal tract includes nasal

obstruction, mass in the nasal cavity, rhinorrhea and epistaxis as seen in our case(9,15). Approximately ,18% of the patients presents with metastasis at the time of diagnosis .The most common site of distant metastasis are bones and lungs.

In sino-nasal tract,the radiological differentials include rhabdomyosarcoma ,lymphoma,poorly differentiated carcinomas and melanoma (10,12,13).Another potential differential in adolescent male could be of juvenile angiofibroma due to recurrent epistaxis. However, it can be ruled out by lack of blush of contrast in digital subtraction angiography in the maxillary artery which is the main artery responsible for recurrent epistaxis in cases of juvenile angiofibroma .

The definitive diagnosis of Ewings sarcoma of the sinonasal tract can be made only on histopathological features and confirmed by immunohistochemical expression of CD99 and NKX2.2(8,14). Embryonal rhabdomyosarcoma is ruled out by absence of cambium layer on histopathology and lack of immunoreactivity with desmin.Non Hodgkin lymphoma, another morphologic differential, is ruled out by the presence of pseudo rosettes and absence of immunoreactivity with CD45. Most cases also show translocation of the Ewings sarcoma gene (EWS) which is located on 22q12 .EWS gene is fused with friend leukemia virus integration site 1 gene(FLI-1) ,which is located on 11q24(8,9).

The prognostic factors include the site of primary tumor,age and presence of distant metastasis (10,11).The successful treatment of the Ewings sarcoma of sinonasal tract includes a multilevel approach including surgery followed by adjuvant radiotherapy and chemotherapy (10).The recommended chemotherapy regimen is alternating cycles of vincristine-doxarubcin-cycophosphamide and ifosfamide-etopside(6) .In our case the patient underwent surgical excision of the mass followed by chemotherapy. The patient is currently doing well and is undergoing chemotherapy with resolution of presenting symptoms and no new complaints.

CONCLUSION

Ewing's Sarcoma originating from sinonasal tract is a rare entity .It is seen mainly in adolescent males. The main stay of diagnosis is histopathology coupled with immunohisto-chemistry and extent can be delineated by contrast enhanced CT neck. The mainstay of treatment is surgery followed by chemo-therapy and radiotherapy.

DECLARATION

Ethics approval and consent to participate: No ethical approval is required

Author's contribution: All the authors contributed to the study conception and design.

Competing interests: The authors declare that they have no competing interests

Informed consent: Informed consent was obtained from the participant and parents included in the study.

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ABBREVIATION

CECT - Contrast Enhanced Computed Tomography
OMC - Osteomeatal Complex
EFST - Ewings family of Skeleton Tumours
AP - Anteriorposterior
TR - Transverse
CC - Craniocaudal
EWS - Ewings sarcoma gene

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