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A rare case Ewing's sarcoma presenting as nasal mass

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Introduction

Ewing's sarcoma is a highly malignant, small round cell tumor that arises from the primitive neuroectodermal cells.[1-2] This was first described by an American pathologist in 1921.[2-3] They are the most common malignant bone tumor in children after osteosarcoma.[4-6] It mainly affects the bones and soft tissues in children and young adults.[7-9] The most common bones affected are pelvis, femur, humerus, ribs and clavicle.[7,9-10] Primary location in head and neck region is uncommon and accounts for 1-4% of all Ewing's sarcomas.[4,11] Primary sino-nasal location is even rarer. Only 14 cases of Ewing's sarcoma involving the nasal cavity or paranasal sinuses have been reported in the literature.[1,3,12-13] About 80% of the incidences are seen in the second decade of life with a male preponderance. In the present article, we present a case of Ewing's sarcoma of right maxillary presenting as bilateral nasal polyps.

Case Report

An 18 year old female presented with complaints of progressive nasal obstruction and epistaxis since 4 months. On endoscopic examination, bilateral highly vascular nasal polyps were found which were adherent to the nasal septum and turbinates. Multiple detector computed tomography (MDCT) revealed bilateral large hypervascular polypoidal masses arising from the right maxillary sinus measuring 17x9 mm. The polyps of the nasal cavity measured 40x27 mm and were adherent to the nasal septum and turbinates. Left maxillary sinus was normal. There was no bony erosions or contiguous spread, other sinuses, cranial fossa and orbit were uninvolved following which an excision biopsy was done for histopathological examination.

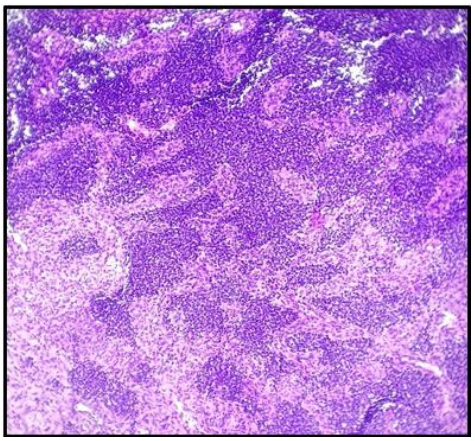

Fig 1: Undifferentiated tumor with dark and light areas. H&E, x4

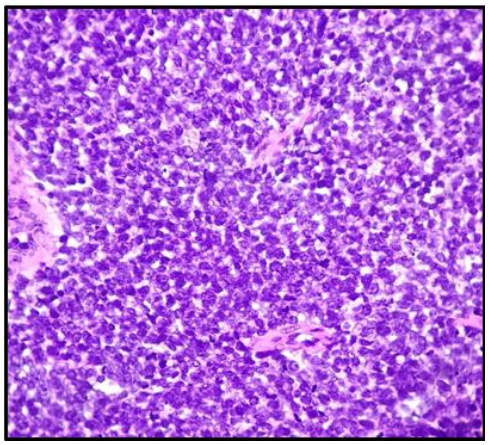

Fig 2: Small round blue cells with fine chromatin. H&E, x40

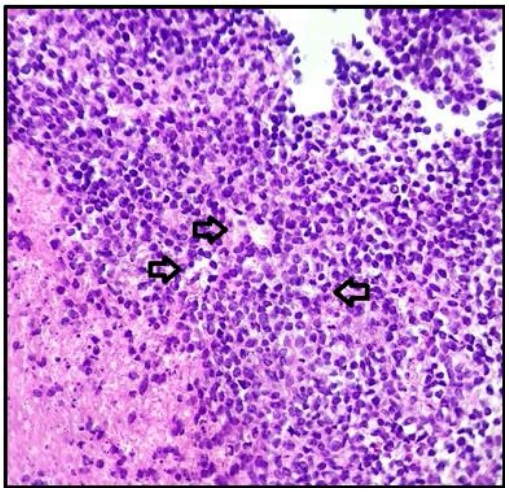

Fig 3: Rosettes marked with black arrows. H&E, x40

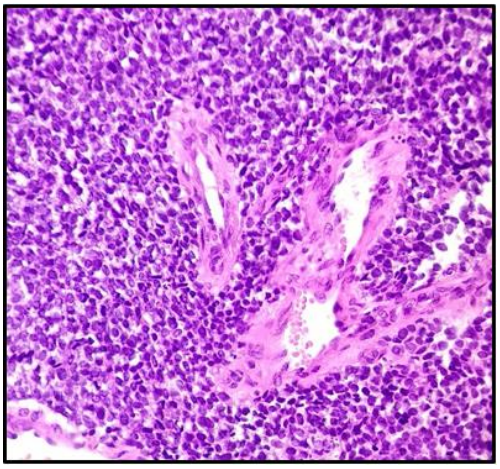

Fig 4: Blood vessels with sheets of tumor cells. H&E, x40

On gross examination, multiple grey white to grey brown tissue fragments were received. Microscopic examination with hematoxylin and eosin (H&E) stain revealed an undifferentiated malignant tumor composed of solid sheets and nests of small round blue cells with fine chromatin, indistinct cell membranes, scant cytoplasm, few rosettes and atypical mitoses. The differential diagnoses of small round blue cell tumors like Ewing's sarcoma/ Primitive neuroectodermal tumor (PNET), olfactory neuroblastoma, Embryonal rhabdomyoscarcoma, lymphoma, neuroendocrine carcinoma were considered. Immunohistochemically, the tumor cells were strongly positive for FLI-1 and were negative for CD45 and desmin. The diagnosis of Ewing's sarcoma /PNET was confirmed.

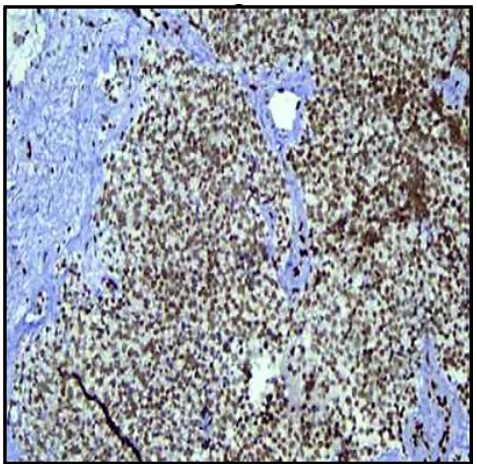

Fig 5: Strong immunopositivity for FLI-1, x10

Discussion

Ewing's sarcoma is a highly malignant small round cell tumor originating from the neuroectodermal cells. Primary Ewing's sarcoma occurs in early childhood or adolescence and rarely in adulthood with a slight male predominance of 1.5:1 and are common in white population.[1,3,4,14-15] Tumors arising from nasal cavities and paranasal sinuses present with non-specific symptoms like nasal obstruction, rhinorrhea and epistaxis. Therefore, they are locally advanced or even metastatic at the time of diagnosis.[4,10,16-17] Majority of the patients have t (11;22) (q24;q12), this fusion protein blocks the differentiation of pluripotent marrow stromal cells. Rest 10-15% of the cases have t(21;22)(q22;q12). In less than 1% of cases, t (7;22), t (17;22), t (2;22) and inv (22) have been found. Mutations associated with P53 or P16/p14 ARF have high aggressive behavior and poor chemotherapeutic response.[3,13]

The differential diagnosis of Ewing's sarcoma in nasal cavity and sino-nasal mucosa includes olfactory neuroblastoma, embryonal rhabdomyosarcoma, lymphoma and neuroendocrine carcinoma. The CT scan would be the radiographic study of choice and the most common radiologic sign in osseous tissue sarcomas are expansion/erosion of the cortical bones.[2] The diagnosis of Ewing's sarcoma/PNET is made with tumors displaying the typical histomorphological features along with supporting immunohistochemical and/or molecular findings. The morphological features include small round blue cells with fine

chromatin and indisctinct scant cytoplasm, occasional Homer-Wright or Flexner-Wintersteiner rosettes. In the present case, the immunohistochemical strong positivity for FLI-1, absence of CD45 and desmin were consistent with the diagnosis of Ewing's sarcoma/PNET.

Conclusion

Ewing's sarcoma/PNET of sino-nasal tract is a rare entity. Radiological imaging like MDCT and immunohistochemistry would be of help in differentiating it from other small round blue cell tumors. Although prognosis may be ominous, it would help in early diagnosis and appropriate management.

Source of Funding

None.

Conflict of Interest

None.

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