



Journal Homepage: -www.journalijar.com
**INTERNATIONAL JOURNAL OF
 ADVANCED RESEARCH (IJAR)**

Article DOI:10.21474/IJAR01/5739
 DOI URL: <http://dx.doi.org/10.21474/IJAR01/5739>



RESEARCH ARTICLE

PERIPHERAL PRIMITIVE NEUROECTODERMAL TUMOUR AS A RARE DIFFERENTIAL IN THE DIAGNOSIS OF SKIN MALIGNANCIES.

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Manuscript Info

Manuscript History

Received: 01 September 2017
 Final Accepted: 03 October 2017
 Published: November 2017

Key words:-

primitiveneuroectodermaltumours, skin, rare.

Abstract

Peripheral primitive neuroectodermal tumours are a group of rare sarcomas arising from the pluripotent neural crest cells. They share similar histology and cytogenetics with Ewings sarcoma. Occurrence of peripheral PNET of the skin is a rare entity and poses challenge with respect to diagnosis and treatment. We report this case to highlight the importance of IHC and cytogenetics in diagnosing skin PNETs which is crucial for its prompt treatment.

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Case:-

A 35 years old female patient presented to our department with recurrent swelling of the left cheek since an year. She gave a history of excision of the lesion been done in her native place 10 months before, the histopathology of which was suspicious of Amelanotic melanoma. She developed recurrence in the next 2 months, and came to us with a lesion of 6x8 cm size, extending from the medial canthus of left eye to left ala of the nose, laterally upto the zygomatic prominence. It was hard on palpation and slightly tender. No neck nodes were palpable. No other lesion or lump were seen elsewhere in the body. Patient's general condition was good and the rest of the parameters were within normal limits.

Computed Tomography of the face and neck were done that showed gross ill defined dermal and epidermal soft tissue thickening in the left pre maxillary region extending into medial canthus, in the nasal bridge on the left side abutting the left nasal bone and into the left inferior eyelid in the subcutaneous plane with no intracranial extension, no intra orbital extension. No underlying bone destruction. Her chest x-ray was normal. Computed tomography of whole abdomen and her bone scan showed no evidence of metastasis.

Henceforth patient was planned for surgery and a wide local excision of the lesion with 5 mm margin along with left superficial parotidectomy and left anterior neck dissection was done. Her facial defect was reconstructed with a forehead flap.

The histopathological report showed features of Malignant round cell tumour arranged in diffuse pattern and exhibiting focal rosettes. All margins and base were free of tumour. Out of the 35 lymph nodes that were removed

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15 were positive for metastasis. The tissue was sent for IHC that showed strong membranous positivity for CD99 (the glycoprotein for MIC-2 antigen), FLI-1, BCL-2, and CD 10. Also VIMENTIN, CD-56 and NSE showed weak to moderate positivity in many tumour cells. This confirmed the diagnosis of peripheral PNET and thus our patient was subjected to combination Chemotherapy based on the institutional protocol for Ewings family of tumours (EFT)2001. Presently patient was put on maintenance chemotherapy, the drugs included vincristine, ifosamide, etoposide, cyclophosphamide, doxorubicin and actinomycin. After 7 months of starting of chemotherapy she developed recurrence lesions on the left eyelid and upper neck. Hence she is on second line chemotherapy now.

Discussion:-

Cancers of the skin are broadly divided as Melanomas and Non-melanoma skin cancers.¹ Among the non melanoma skin cancers, Basal cell carcinoma is the commonest worldwide accounting for almost 70%.² Squamous cell carcinomas occupy most of the remaining except for a very small percentage contributed by cancers like dermatosarcoma protuberans, merkel cell carcinomas, pagets disease of skin, kaposissarcoma, angiosarcoma etc.^{2,3} Primitive neuroectodermaltumour involving skin as a primary organ is very less often seen and not more than 50 cases have been reported till date.⁴ The under-reported in literature is also due to its aggressive behavior and confusion regarding the diagnosis. Based on the site of origin, PNETs are classified as central (brain and spinal cord) and peripheral PNETs.⁵ Peripheral PNETs have considerable overlap with the Ewings sarcoma of bone both sharing a common translocation t(11,22)(q24,q12) fusion gene designated as (EWS/FLI-1).⁶ Peripheral PNETs occur in various locations like Askins tumour of the thoraco-pulmonary region, maxillofacial and eye PNETs, peripheral limbs, intra abdominal and even gynaecological organs accounting for nearly 1% of all sarcomas.⁷ Eventhough commoner in children PNETs are also found in adults with a mean age between 5 to 77 years. However 80% of tumours occur in less than 15 years.⁸

On the levels of Histopathology, PNETs are highly cellular, with small round blue cells arranged usually as sheets and with pseudo rosettes. Other tumours with similar appearance include rabdomyosarcoma, neuroblastoma and non hodgkinslymphoma, thus cannot be distinguished solely based on histological grounds. Electron microscopy may show neurosecretory granules with microtubules and microfilaments. Short dendritic processes lie between the cells of peripheral PNETs unlike Ewings sarcoma where such dendritic processes are absent.

Elucidation of IHC is necessary to distinguish PNETs with other small round cell tumours. The expression of MIC-2 produces an antigen MIC-2 which is consistently identifies Peripheral PNETs as well as Ewings sarcomas. However CNS PNETs lack MIC-2 expression. CD99 (the glycoprotein for MIC-2) and vimentin co-expression is typical to PNETs.⁹ Other markers like S100, Neuron specific enolase, CD75 and synaptophysin are however non specific.

CT scan or Magnetic resonance imaging of the skin PNETs is essential to know the extent of the disease as well as to rule out the involvement of underlying bone. PNETs have a high incidence of metastatic disease at presentation, therefore a full metastatic workup as CT scan of the chest and abdomen, bone marrow biopsy and technicium 99 bone scan are mandatory.

Treatment of the skin PNETs is a multimodality approach and depends on whether the disease is localized or metastatic. Complete resection with negative margins wherever possible prognosticates better survival. Post operative adjuvant chemotherapy has proved be beneficial with regimens recommended including drugs like vincristine, doxorubicin, cyclophosphamide, with ifosomide and etoposide.¹⁰ The treatment of metastatic disease includes neo adjuvant or adjuvant chemotherapy with radiotherapy to all the sites of gross disease. Surgical excision may be an option where possible.

Conclusion:-

Skin PNETs are as much rare as they are aggressive. Their identification and documentation is essential in all clinicopathologically suspicious cases. Prompt confirmation of the diagnosis using the ancillary techniques like IHC and molecular analysis would greatly aid in the accurate treatment of the disease and modify the outcome for towards better prognosis.



Figure 1:-pre- operative picture of the patient.



Figure 2:-post operative photograph of the patient.

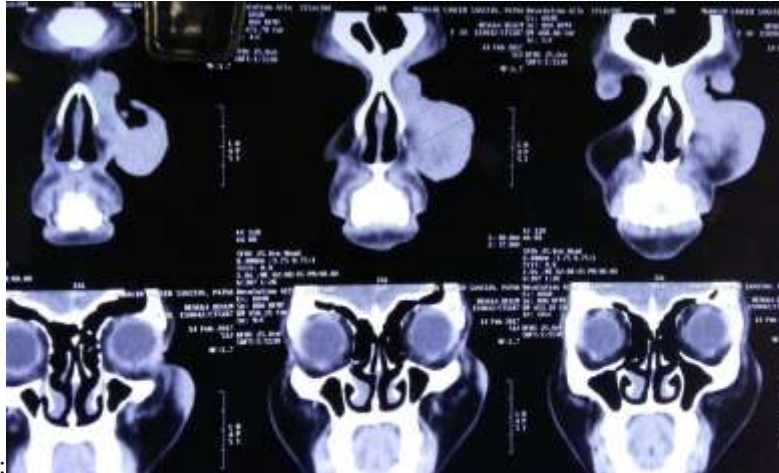


Figure 3:-ct scan image showing soft tissue extension of the tumour.

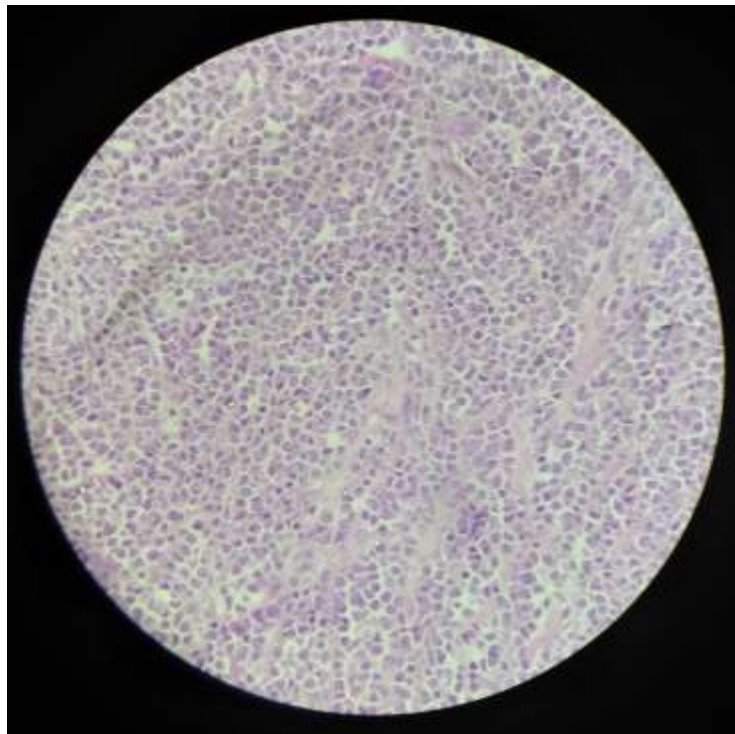


Figure 4:- H&E 40x histopathology



Figure5:- Recurrence Left Eyelid And Upper Neck

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