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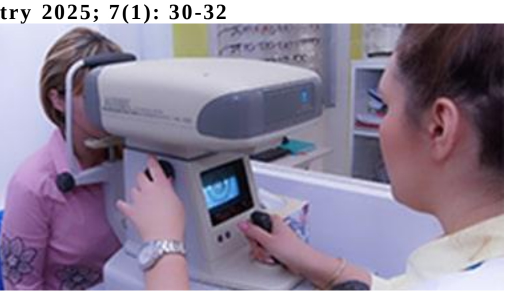

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A rare case of primitive neuroectodermal tumor (PNET) of conjunctival localization Un cas rare de tumeur neuroectodermique primitive (PNET) de localisation conjonctivale

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Abstract
Summary: PNETs (primitive neuroectodermal tumors) are a group of tumors, highly malignant with high metastatic potential. They are classified into two categories: central PNETs and peripheral PNETs. Conjunctival localization is extremely rare and poses diagnostic problems; few cases have been reported in the literature. Biopsy and histopathological analysis are the key to diagnosis.
We report the case of a peripheral PNET with conjunctival localization in a young 24-year-old patient.
Résumé: Les PNET (tumeurs neuro-ectodermiques primitives) forment un groupe de tumeurs, hautement malignes avec un potentiel métastatique élevé. Elles sont classées en deux catégories: les PNET centraux et les PNET périphériques.
La localisation conjonctivale est extrêmement rare et pose des problèmes diagnostiques, peu de cas ont été rapporté dans la littérature. La biopsie et l'analyse histopathologique sont la clé du diagnostic.
Nous rapportons le cas d'une PNET périphérique à localisation conjonctivale chez une jeune patiente de 24 ans.

Keywords: Primitive neuroectodermal tumors histopathological analysis, case report

Introduction
PNETs (primitive neuro-ectodermal tumors) are a group of small, round, highly malignant tumors derived from primitive neuro-ectodermal progenitor cells. They are classified into two categories: central PNETs and peripheral PNETs.
Peripheral PNETs are mainly seen in bone and soft tissue in children and young adults, primary visceral locations have also been reported, such as the kidney, uterus, ovary, gallbladder, parotid gland, pancreas and testis [2]. The best-known forms are Ewing's sarcoma and Askin's tumour [1].
Conjunctival localization is extremely rare. To date, only 4 cases have been reported in the literature.
We report the exceptional observation of a conjunctival PNET in a 24-year-old patient in order to raise diagnostic difficulties.

Observation
A 24-year-old woman with no particular pathological history presented with a conjunctival lesion of the right eye that had been present for three months and was gradually increasing in size (Figure 1).
The initial ophthalmological examination revealed a round, brownish-red, soft, well-circumscribed, temporal para-limbic bulbar conjunctival lesion measuring between 0.5 and 2 cm in long axis.
This patient underwent an excisional biopsy with mitomicine C treatment.
The anatomopathological study revealed a tumor proliferation with round cells, lobulated architecture and uniform appearance. The tumor cells had ovoid nuclei with fine granular chromatin and small nucleoli with sparse cytoplasm compatible with the diagnosis of primitive neuro-ectodermal tumor of the conjunctiva (Figure 2).

Fig 1: Brownish conjunctival growth in our 24-year-old patient

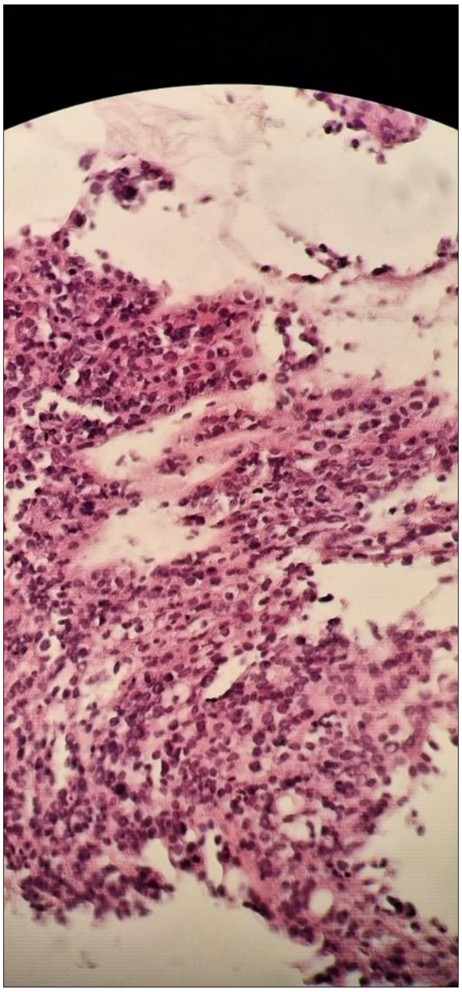

Fig 2: histological appearance of the tumor at high magnification (x20), showing cells with ovoid nuclei, fine granular chromatin, small nucleoli and reduced cytoplasm

A total resection of the tumor was performed in this patient. The extension study revealed no secondary localizations.
The patient was followed for a period of 24 months without any sign of local tumor recurrence or systemic metastasis.

Discussion

PNETs (primitive neuroectodermal tumors) are a group of highly malignant, small round cell tumors derived from primitive neuro-ectodermal progenitor cells. They are classified into two categories: central PNETs and peripheral PNETs. Batskis *et al* classified PNETs into three categories based on tissue of origin: central nervous system PNETs (tumors derived from the central nervous system), neuroblastoma (tumors derived from the autonomic nervous system), and peripheral PNETs (tumors derived from tissues outside the central and autonomic nervous systems) [1].

PNETs have an equal predisposition according to gender and a worse prognosis in the older age group [4].

These very rare tumors share a similarity of biochemical and oncogenic expression with several other tumors, such as Askin's tumor or Ewing's sarcoma, which makes them belong to the family of Ewing's sarcomatous tumors which all derive from the same stem cell [6].

pPNET has been reported in various locations including the orbit and conjunctiva and has been characterized by its aggressiveness and the occurrence of metastasis. A lower tendency for metastasis and a better prognosis for orbital pPNET have been attributed to the presentation of symptoms, thus favoring early medical consultation and prompt management.

The tumor may be solid or cystic. It grows invasively and may be responsible for complications such as hemorrhage or necrosis. The clinical feature is nonspecific and is similar to any other aggressive neoplasm of the conjunctiva. The final diagnosis depends on the histopathological examination which finds small uniform round cells distributed diffusely, sometimes lobulated with areas of focal necrosis [5].

4 cases of PNET with conjunctival localization have been reported: a young woman and three young men who benefited from a complete excision, two of whom had received chemotherapy, no tumor recurrence was diagnosed during the years of follow-up [4, 5, 7, 8].

We report here the fifth case of PNET with conjunctival localization that underwent complete excision and has been in remission for more than 2 years.

The oncologist suggested surgical resection alone and regular surveillance similar to Chow's experience for cutaneous lesions and localized subcutaneous tumors of the Ewing sarcoma family.

Conclusion

Conjunctival PNETs are very rare and aggressive tumors. Given their undifferentiated morphological appearance and their rarity, they can pose diagnostic and therapeutic problems. Biopsy and histopathological analysis are the key to diagnosis.

Human and animal rights

The authors declare that the work described has been carried out in accordance with the Declaration of Helsinki of the World Medical Association revised in 2013 for experiments involving humans as well as in accordance with the EU Directive 2010/63/EU for animal experiments.

Informed consent

Consent was obtained, and the patient was made aware that her medical records would be kept confidential.

Author contributions

All authors attest that they meet the current International Committee of Medical Journal Editors (ICMJE) criteria for Authorship.

Conflict of interest

The authors have no conflicts of interest.

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