Primitive neuroectodermal tumour of the orbit: A case report and literature review

Imane Bekkouch, MD[1], Tayeb Kebdani, MD[1], Ouafae Masbah, MD[1], Issam Lalya, MD[1], Ahmadou Tolba, MD[1], Lamiae Kanouni, MD[1], Khalid Hassouni, MD[1], Noureddine Benjaafar, MD[1], Brahim Khalil El Gueddari, MD[1]

(1) National institute of oncology, Faculty of medicine of Rabat, University Mohammed V, Morocco.

✉ *Corresponding Author: Dr Imane Bekkouch, MD*
National institute of oncology, Faculty of medicine of Rabat, University Mohammed V, Morocco.
E-mail: bekkiman78@yahoo.fr

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Abstract

Primitive neuroectodermal tumours are rare. The orbital localization is exceptional. Only 13 cases were described in the literature; which only three are adults. We report a case of a 59-year-old man, who was admitted for exophthalmia, NMR revealed a tumour in the left orbit. Pathological examination and Immunohistochemical studies showed a Primitive neuroectodermal tumour. The patient was treated with chemotherapy followed by radiation therapy.
After 22 months of follow-up, the patient remained free of disease

Introduction

Primitive neuroectodermal tumour (PNET) is a term used to describe a category of highly Malignant small round cell tumours of neuro-ectodermal origin with variable cell differentiation [1, 2]; primitive neuroectodermal tumours are rare, they represent less than 1 % of all the sarcomas.
The majority of PNET occurs in the central nervous system (SNC).
PNET recognized outside of the CNS are diagnosed as peripheral PNET. The orbital localization is exceptional only 13 cases have been reported previously; we present the clinical, radiological and histopathological features of an orbital mass in a 59 year-old man.

Observation

A 59-year-old man presented with a 10-month history of exophthalmia.
Nuclear magnetic resonance NMR revealed a heterogeneous hyper intense signal on a T2-weighted image in the left orbit.
Pathological examination showed a sheet of small cells with irregular nuclei. Immunohistochemical studies demonstrated positive immunoreactivity for neurone-specific enolase and synaptophysin, On the basis of these findings, a diagnosis of primitive neuroectodermal tumour of the orbit was made. The patient was treated with three courses of chemotherapy consisting of, ifosfamide, etoposid doxorubicin and cysplatin, the response was estimated at 50 % by NMR. Then he was treated with 60 Gray of external beam radiation therapy (2Gray per fraction and 30 fractions). After 22 months of follow-up, the patient remained free of disease and a repeat NMR was normalized, with no sign of residual tumour.

Discussion

Primitive neuroectodermal tumours are very uncommon.
The orbital localization is exceptional; only 13 cases have been reported in the literature [1, 3].
The value of this reported case is related to his age, since all reported cases in the literature are pediatrics, except 3 patients, and all the three are man. (Table 1).

Table 1: table including all reported cases in the literature starting from 1986

Authors name	Case age	Case sex	Year of publication	Reference
S Shuangshoti and al	52-year-old	man	1986	Br J Ophthalmol. 1986 July; 70(7): 543–548.
Wilson WB and al	7-year-old	girl	1988	Cancer. 1988 Dec 15;62(12):2595-601
Singh AD and al	10-year-old	girl	1994	Arch Ophthalmol. 1994 Feb;112(2):217-21.
Bansal RK and al	8-month-old	boy	1995	Indian J Ophthalmol 1995;43:29–31.
Kiratli and al	28-year-old	man	1999	Ophthalmology. 1999 Jan; 106(1):98-102.
Alyahya GA and al	5-year-old	girl	2000	Graefes Arch Clin Exp Ophthalmol. 2000 Sep;238(9):801-6.

Authors name	Case age	Case sex	Year of publication	Reference
Sen S and al			2002	J Pediatr Ophthalmol Strabismus. 2002 Jul-Aug;39(4):242-4.
Lezrek M and al	13-year-old	Boy	2005	J Fr Ophtalmol 2005; 28: e8.
Romero R and al	6-year-old	Boy	2006	Arch Soc Esp Oftalmol. 2006 Oct; 81(10):599-602.
Znati K and al	23 year-old	man	2006	Revue Française des Laboratoires, Volume 2006, 380, March 2006, 41-43.
Tamer C and al	10-year-old	boy	2007	Can J Ophthalmol. 2007 Feb;42(1):138-40
Romero IL and al	10-month-old	girl	2008	Arq Bras Oftalmol. 2008 Nov-Dec;71(6):871-3.
Kim UK and al	2 year old	girl	2009	Indian ophtal J; 10; 2009; 200-202.

The diagnosis is always based on histological analysis and immunohistochemical staining with or without cytogenetically study.
Histologically, PNETs are highly cellular and demonstrate a monotonous pattern of small round cells with hyper chromatic nuclei, and high nuclear–cytoplasm ratio with varying degrees of neuronal differentiation. This progressive process begins with neuron specific enolase (NSE) expressivity, followed by Homer–Wright rosette formation, phenotypic ganglion cell differentiation, and finally by neurofilament protein expression [4, 5].

The differential diagnosis of PNET of the orbit includes the other small blue round cell tumours: lymphoma neuroblastoma, Ewing's sarcoma, rhabdomyosarcoma, small cell osteogenic sarcoma, and mesenchymal chondrosarcoma [1.6].
There is no established treatment protocol for patients with orbital PNET ,these tumours progress rapidly and often have already metastasized at the time of diagnosis [7].Because of the close relationship with Ewing Sarcoma, supplementary chemotherapy, radiation therapy or both is currently recommended [8,9.10].

These tumours prognosis is poor; their evolution is marked by local recurrences and metastasis. Survival ranged from 1 month to 5 years. For 6 cases of 13 cases of orbital PNET report in the literature, the survival was between 6 and 45 months.

Conclusion

Primitive neuroectodermal tumour of the orbit is extremely rare, especially in adults. the differential diagnosis of PNET of the orbit includes the other small blue round cell tumours. Treatment is based on chemotherapy associated to radiation therapy or surgery. Prognosis is very poor.

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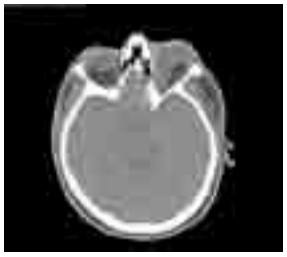
Figures

A computed tomography scan showed a left orbital mass



Radiotherapy by 2 Fields, anterior and left side Field.