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Primary Brain Ewing Sarcoma/pPNET in Elder Adult

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Abstract

The Ewing sarcoma/peripheral primitive neuroectodermal tumour type tumour (EWS/pPNET) group includes those small round blue cell tumours with morphological attributes of the germinal neuroepithelium. It represents a group of rare primary intracranial tumours, mostly described in paediatric population. There are very rare reports of primary intracranial EWS / pPNET in elders. We report the case of an 83-year-old man, where a cerebellar-pontine lesion mimicking a meningioma turned out to be a EWS/pPNET. Genetic studies were not performed, as he was not to be submitted to chemotherapy.

Keywords: EWS/pPNET; Ewing sarcoma; Peripheral primitive neuroectodermal tumour; Cerebellar-pontine angle

Introduction

Ewing sarcoma / peripheral primitive neuroectodermal tumour type tumour (EWS / pPNET) are very rare tumours (1% of all sarcomas); Ewing's sarcoma represents 5 to 15% of malignant bone and soft tissue tumours; two thirds of cases of Ewing's tumours occur before age 35 years, with a median age of 20 years [1]. Primary brain tumour cases in adults are found in reports literature [2,3]. Ewing sarcoma / peripheral primitive neuro ectodermal tumour type tumour (EWS / pPNET), belongs to a tumour family that shares clinicopathologic and molecular genetics features. Histologically, its family is recognized as a small, round, blue cell tumour, staining positively for CD99 and usual genetic features gene rearrangements between chromosome 22 and 11 (22q11 *EWSR1/FLI1*) or *FUS* (chromosome 16) [4]. Primary brain tumours of this sort are rare (a review of the literature revealed 19 cases), being the eldest a 67-year old female [5-22]. They are classified per the 2016 WHO Classification as mesenchymal, non-meningothelial tumours [4].

Review of the Case

We report the case of an 83-year-old man, autonomous, who presented at the emergency department with complaints of numbness on the right side of the face lasting 3 months. The patient underwent a head-CT, which demonstrated the presence of an extra-axial lesion on the right cerebellar-pontine angle, approximately 2cm wide, and was referred for a neurosurgical appointment. He returned a month later to the emergency department complaining of a severe loss of balance that deemed him unable to walk. He repeated the head-scan which showed that the lesion had doubled in size within this timespan. A head-MRI was performed and the lesion resembled a meningioma. The patient underwent surgery 2 weeks later and total removal of the lesion was achieved via retrosigmoid craniotomy. In the immediate postoperative period, he presented a right peripheral facial palsy, House-Brackman 4, and therefore a tarsorrhaphy was performed. He recovered partially of his facial palsy, and is now able to walk. No further lesions were found and he is scheduled to start radiotherapy [23].

Discussion

The histology of the lesion revealed a small, round cell tumour with an immunophenotype favouring the Ewing sarcoma / peripheral primitive neuroectodermal tumour type tumour (EWS / pPNET). Genetic studies were not performed in this case, since the patient was not to be submitted to chemotherapy. These are very rare tumours, mostly appearing on paediatric age and therefore the diagnosis and prognosis factors are unknown. Ibrahim et al. [24] in their recent review, proposed some possible prognostic factors, some of which are applicable to Ewing sarcoma in a broader sense. Specifically, age greater than 17 years, inaccessibility of the tumour for surgical resection, incomplete resection, multifocality, and tumour genetic factors (e.g. Type 1 fusion gene) appear to have negative prognostic implications. There is not enough data on the matter to draw a

conclusion other than it is a diagnostical challenge to distinguish these tumours from other with similar characteristics; but it is fundamental to do so, ensuring that proper follow-up and complementary treatments are administered.

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