Temporal primary cerebral Ewing sarcoma extended to skull

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Cerebral Ewing´s sarcoma is a very rare disease, few cases have been reported on literature[1,2]. Any bone can be affected, some rare intracranial primary cases had been reported[3], The peak incidence is between 10 and 20 years old. Males are more frequently affected than females, and usually presents as a solitary bony lesion[1-3]. Clinically the most important and earliest symptom is pain, which is initially intermittent but becomes very intense[1-3].

CASE

A 30 year old female with a history of tonic-clonic generalized seizures and aphasia which started at 8 years old. She presented seizures characterized by loss of consciousness and tonic-clonic movements. Neurological examination showed absent corneal and nasal reflexes, right facial, central paresia, right hemiplegia, and abnormal Babinski. The MRI showed a lesion enhancement tumor with low signal heterogeneous gadolinium enhancement, whereas signal intensity on T2-weighted images varies after contrast enhancement (Fig 1A). The spectroscopy showed an increase of coline and important diminution of N-acetil-aspartate (Fig 1B).

The patient underwent left frontal craniotomy and the tumor was excised totally. Following surgery, she underwent whole brain, spine and local radiation therapy (30 Gy in total) and also received chemotherapy. She died 11 days after the surgery.

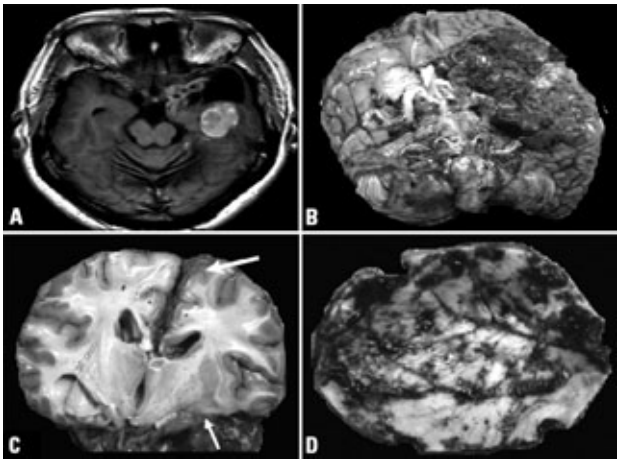

Fig 1. [A] The MRI-imaging showed a lesion signal intensity on T2-weighted images varies after contrast enhancement. [B] Brain gross aspect. [C] The tumor was an irregular tan-gray mass of 4×4 cm, on the temporal and parietal right lobes. On a surface cut, focal areas of glistening grayish-white substance were admixed with poorly demarcated firm areas, necrosis and hemorrhage and in [D] showed dissemination along skull base.

Partial autopsy was performed, grossly; the tumor was an irregular glistening gray mass of 4×4 cm, which extended all along the temporal and parietal bone on the right side of the skull (Fig 1C). Tumor surfaced poorly demarcated firm areas, with necrosis, hemorrhage and dissemination along the skull base (Fig 1D).

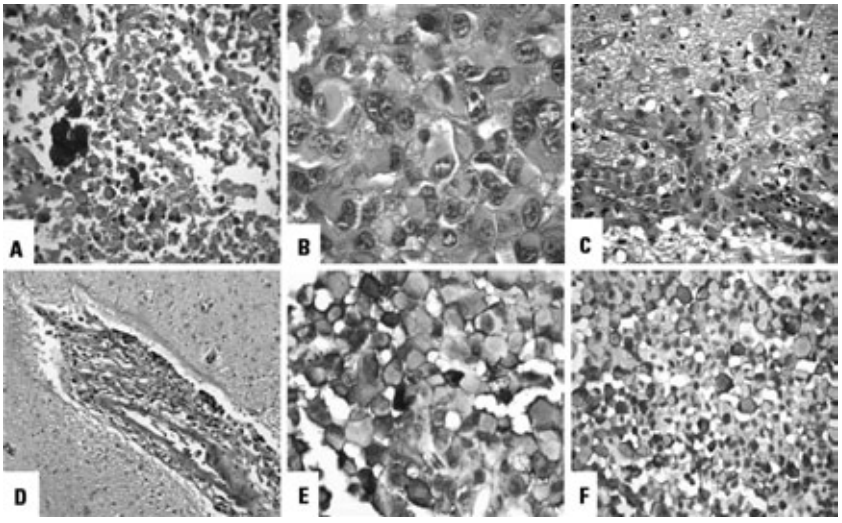

Fig 2. Histological features. [A] Showed a tumor composed of bland spindle-shaped cells with indistinct pale eosinophilic cytoplasm and small hyper chromatic oval nuclei. [B] In the cellular areas, the cells showed nuclear polymorphism with high levels of mitotic activity and ring cells were observed also. [C] The tumor cells were embedded in a variably fibrous or myxoid stroma that tended to alternate in different areas of the tumor. [D] A prominent network of branching capillary-size blood vessels was also seen. Reactive gliosis was observed in the adjacent brain parenchyma. [E] Immunohistochemistry, Tumor was CD99 immunoreaction cells and [F] CD117 positive immunoreactions in the most of the neoplastic cells (IHQ ×400).

Histologycally tumor revealed bland spindle-shaped cells with indistinct pale eosinophilic cytoplasm and small hyperchromatic oval nuclei, ring cell likes were also observed (Fig 2A and 2B), ploemorphism and mitotic features (Fig 2C and 2D). The tumor cells were embedded in a variably fibrous or myxoid stroma that tended to alternate in different areas of the tumor. Immunohistochemical analysis of the tumor cells revealed diffuse expression of vimentin, CD99 (Fig 2E) and CD117 (Fig 2F). Ewing sarcoma was diagnosed. The tumor was resistant to different kinds of therapy.

DISCUSSION

Primitive neuroectodermal tumors are in the Ewing's sarcoma family of tumors and are composed of small round cells, belong to a family of tumors that share clinic pathologic and molecular genetic features, including the characteristic chromosomal translocation that results in the fusion of the EWS gene on 22q12 to either the FLI1 gene on 11q24 or other[2-4]. One basic distinction is between primitive neuroectodermal tumors of the central nervous system (cPNETs) and primitive neuroectodermal tumors of the peripheral nervous system (pPNETs), which are clinicopathologically and genetically distinct[3,4]. Among the cPNETs including medulloblastoma, pineoblastoma, cerebral neuroblastoma, ependymoblastoma, medulloepithelioma, primary rhabdomyosarcoma, and atypical teratoid/rhabdoid tumor, whereas the pPNETs comprise the more differentiated end of a spectrum of neoplasms that include skeletal and extraskeletal Ewing's sarcoma. In most instances these entities may be differentiated by a panel of antibodies that should include those with low and high molecular weight cytokeratins, epithelial membrane antigen, type IV collagen, ENE, sinatophysin, CD99, CD56, and S-100 protein[3].

Appropriate treatment is based on a correct diagnosis, the surgical pathologist must be familiar both with basic characteristics of each of the numerous entities as well as the spectrum of morphologic features that each may display[3,4].

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SARCOMA DE EWING TEMPORAL CEREBRAL PRIMÁRIO COM EXTENSÃO PARA O CRÂNIO

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Isolated hypoglossal nerve palsy

An unusual rare presentation in systemic lupus erythematosus

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Systemic lupus erythematosus (SLE) is an immune-mediated disease of unknown etiology which can damage the peripheral and central nervous system[1]. Cranial nerves have rarely been involved in SLE's patients[2,3]. Isolated hypoglossal nerve palsy (HNP) was even rarer reported in patient with SLE[4].

We report a patient with SLE who presented with isolated HNP.

CASE

A 27-year-old woman presented with fever, alopecia, skin rash, photosensitivity, Raynaud's phenomenon, ar-