Article published online: 2023-08-22

https://doi.org/10.1590/0004-282X20180058

A recurrent laryngeal nerve malignant peripheral nerve sheath tumor in a child with neurofibromatosis type 1

Tumor maligno de bainha de nervo periférico acometendo o nervo laríngeo recorrente em uma criança com neurofibromatose tipo 1

Sérgio Ferreira Alves Júnior[1], Leonardo Hoehl Carneiro[1], Bruno Niemeyer[1], Elise Tonomura[1], Edson Marchiori[1]

An 11-year-old girl presented with hoarseness, dysphagia, and swelling in the left side of her neck. She had a previous diagnosis of neurofibromatosis type 1, presenting with multiple café-au-lait spots and subcutaneous neurofibromas (Figure A). Magnetic resonance imaging showed a mass in the left side of her neck (Figure B and C). The final diagnosis was a malignant peripheral nerve sheath tumor (Figure D) arising from the left recurrent laryngeal nerve. A malignant peripheral nerve sheath tumor is an uncommon, aggressive soft tissue sarcoma of neural origin. It is defined as a nerve sheath tumor arising from a peripheral nerve, a pre-existing peripheral nerve sheath tumor, or in the setting of neurofibromatosis type 1[1,2].

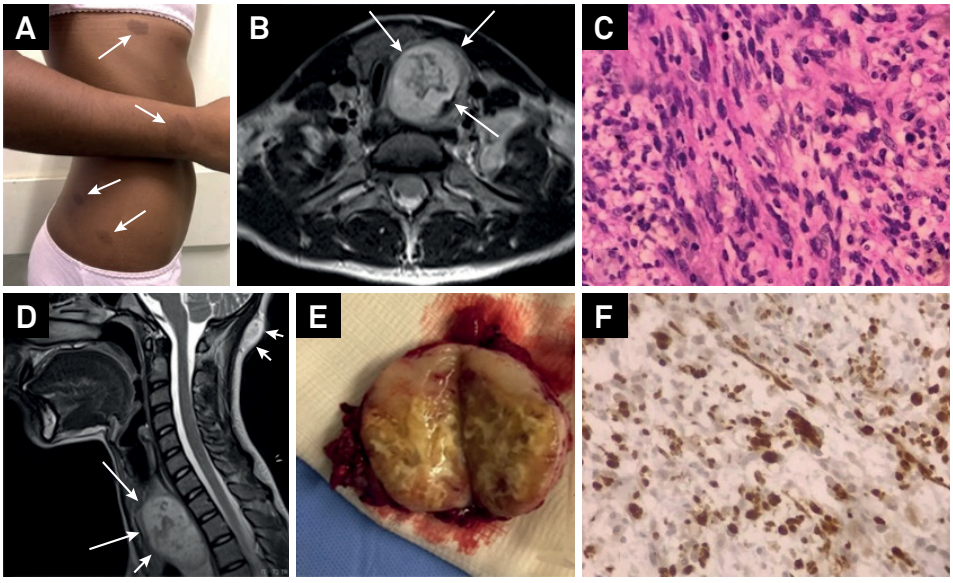

Figure. Ectoscopic examination (A) demonstrated café-au-lait spots on the skin surface. Axial (B) and sagittal (C) T2-weighted magnetic resonance images show a round necrotic mass located at the posterior margin of the left thyroid lobe, along the recurrent laryngeal nerve pathway (arrows). The tumor shows heterogeneous high signal intensity, with a bright center of irregular margin necrosis. The mass displaces the trachea laterally to the right and the left lobe of the thyroid anteriorly. Small subcutaneous neurofibromas are also present in the occipital region (arrowheads). The gross specimen (D) shows a globular, fleshy, tan-white mass with areas of necrotic degeneration and secondary hemorrhage, reflecting the malignity and aggressive behavior of the neoplasm. Photomicrograph (E) shows areas of high cellular density composed of spindle-shaped cells with nuclear atypia and atypical mitosis, which reveal the malignant feature of the tumor, a spindle cell sarcoma (hematoxylin and eosin stain, ×400). Immunohistochemical image (F) demonstrates nuclear and cytoplasmic positive staining for the S-100 protein (a biological marker of neural crest derivative cells, including Schwann cells; ×400).

References

1. Demir HA, Varan A, Yalçn B, Akyüz C, Kutluk T, Büyükpamukçu M. Malignant peripheral nerve sheath tumors in childhood: 13 cases from a single center. J Pediatr Hematol Oncol. 2012 Apr;34(3):204-7. https://doi.org/10.1097/MPH.0b013e31822d4cef

2. Farid M, Demicco EG, Garcia R, Ahn L, Merola PR, Cioffi A, Maki RG. Malignant peripheral nerve sheath tumors. Oncologist. 2014 Feb;19(2):193-201. https:/doi.org/10.1634/theoncologist.2013-0328

[1] Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brasil.

Correspondence: Edson Marchiori; Rua Thomaz Cameron, 438; 25685-120 Petrópolis RJ, Brasil; E-mail: edmarchiori@gmail.com.

Conflict of interest: There is no conflict of interest to declare.

Received 16 March 2018; Accepted 15 April 2018.