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A rare case of a spontaneous neck hematoma in a patient with type 1 neurofibromatosis

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ABSTRACT

Neurofibromatosis type 1 (NF-1) is a genetic disorder that affects one in 3000 individuals. Although NF-1 notably involves nerves and connective tissue, vascular involvement in large series is estimated to range from 0.4% to 6.4%. Jugular vein involvement in these patients is rare. Spontaneous neck hematomas and hemorrhages are also unusual. We present a case of a NF-1 patient with a spontaneous neck hematoma with possible leakage from the left internal jugular vein, presenting as a lateral neck mass. The fragility of the vein wall and the surrounding tissue led patient to a severe intraoperative bleeding. Pathological examination revealed degenerated neurofibroma which was in contact with or infiltrated the vein wall. ENT and other clinicians should be aware of this potentially fatal entity considering that it may present as a lateral neck mass.

1. Introduction

Neurofibromatosis (NF1) is an autosomal dominant disorder which affects one in 3000 individuals. It mainly affects nerves and other tissues and organs such as eyes, skin, and long bones. Although rare, vascular involvement ranges from 0.4 to 6.4% [1]. Venous system involvement is unusual, while internal jugular vein involvement (particularly aneurysms) reported in the literature is limited [2–5]. Spontaneous neck hematomas and hemorrhages in patients with NF-1 are limited as well [6–10].

2. Case report

A 53-year-old man was admitted to our ear, nose, throat (ENT) department with a 4-week history of a left cervical mass with a sudden expansion in the last days. The mass was soft, mildly tender and non-pulsatile. Clinical evaluation revealed multiple neurofibromas over his trunk, extremities and face with café au lait spots (Fig. 1). The diagnostic criteria for NF-1 were fulfilled as well (Table 1). Hoarseness, signs of tracheal compression or dyspnea were absent. There was no history of trauma. In addition, flexible fiberoptic laryngoscopy examination was normal.

A contrast-enhanced computed tomography demonstrated a lobulated mass 7.5 cm × 3.5 cm × 11 cm located medially to the sternocleidomastoid muscle exerting pressure to the internal jugular vein, without clarifying the relationship between the lesion wall and the internal jugular vein wall (Fig. 2a). Magnetic resonance imaging (MRI) revealed a left cervical

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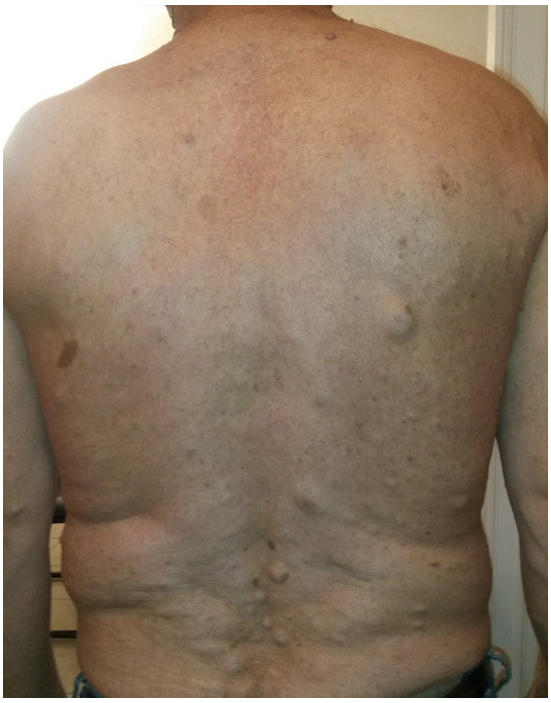

Fig. 1. Clinical presentation of NF-1 with multiple neurofibromas and café au lait macules.

Table 1
Diagnostic criteria for neurofibromatosis type I as defined by National Institutes of Health (NIH). Criteria are met if a patient has two or more of the above features.

(1) Six or more ‘‘cafe’ au lait’’ macules over 5 mm in greatest diameter in pre-pubertal persons and over 15 mm in greatest diameter in post-pubertal persons
(2) Two or more neurofibromas of any type or one plexiform neurofibroma
(3) Freckling in the auxiliary or inguinal regions
(4) Optic glioma
(5) Two or more lisch nodules (iris hamartomas)
(6) A distinctive osseous lesion such as sphenoid dysplasia or thinning of long bone cortex with or without pseudoarthrosis
(7) A first-degree relative (parent, sibling, or offspring) with neurofibromatosis type I, based on the above criteria

heterogenous mass with high signal peripherally on T1-weighted images extending from C2 vertebra to the left supraclavicular fossa, lesion compatible with a hematoma with possible leakage from the left internal jugular vein. Due to extensive bleeding it was not possible to assess the presence of an underlying tumor (Fig. 2b). In addition, digital subtraction angiography and CT angiography revealed thrombosis of the upper left jugular vein (Figs. 3 and 4). At preoperative blood cell tests, blood counts, activated partial thromboplastin and prothrombin time were normal

After obtaining consent and consulting with vascular surgeons the patient underwent surgical exploration. During operation a left jugular vein dilatation was revealed with an organized thrombus and a hematoma around it. The friable vein wall and the surrounding vascular tissue led to severe bleeding adding up to a final total of 1550 ml which was difficult to be controlled, and the patient received three units of packed erythrocytes intraoperatively. The jugular vein, carotid artery and accessory nerve were identified and carefully dissected, in order to properly delineate the tumor limits. Finally, the mass was excised while the jugular vein was ligated and resected along with the tumor (Fig. 5).

As far as pathology is concerned, the paraffin-embedded tissue was cut in 2 μm sections and stained with hematoxylin–eosin, Verhoeff-Van Gieson (VVG) and trichrome Masson stains. The immunohistochemical study performed included a standard avidin-biotin immunoperoxidase technique and S-100 protein (S100) and smooth muscle actin (SMA) antibodies. Histological examination of the excised specimen revealed a hematoma-like structure surrounded partly by disrupted vascular wall elements as shown by histochemistry (VVG) and immunohistochemistry (SMA). The main body showed relative demarcation, organization, an abundance of fibrous connective tissue with presence of small vessels and prominent fibroblastic reaction. The surrounding tissue mainly and parts of the central areas, despite the degenerative changes, presented a diffuse proliferation of wavy, spindle-shaped cells, which immunohistochemically showed focal, but strong S100 expression (Fig. 6). These findings indicated the presence of

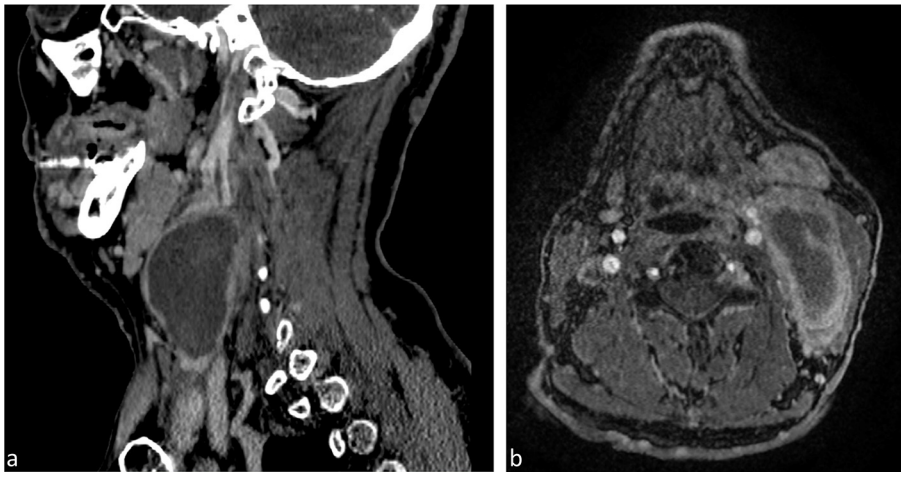

Fig. 2. (a) CT neck scan sagittal plane showing a mass exerting pressure to left internal jugular vein (b) MRI neck scan axial cut. Hematoma with possible leakage from the left internal jugular vein.