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Spontaneous internal carotid artery rupture during neck dissection in osteogenesis imperfecta patient

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ABSTRACT

Objective: Several connective tissue disorders are associated with vascular wall abnormalities, including spontaneous dissection of the cervical arteries. Osteogenesis imperfecta (OI) is a hereditary disorder, with rare neurovascular complications, which potentially lead to life-threatening events.

Methods: We presented a case of spontaneous internal carotid artery (ICA) rupture that occurred in a 52-year-old-woman, suffering from OI type I, and who underwent a bilateral modified radical neck dissection (mRND) plus total thyroidectomy for a T4aN1b thyroid cancer. During mRND, an ICA's spontaneous rupture occurred.

Results: Histopathologic report suggested a structural defect of the arterial wall without cancer infiltration. The patient did not experience any neurologic complications.

Discussion: Head and neck surgeons have to be aware about early clinical recognition of possible cervical vascular abnormalities, in patients with connective tissue disorders, potentially responsible for dramatic vascular rupture during cervical surgical procedures.

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1. Introduction

Many studies report the presence of structural alterations of vascular walls in patients suffering from connective tissue disorders such as Ehlers–Danlos syndrome (eDS), fibromuscular dysplasia, osteogenesis imperfecta (OI) and Marfan syndrome; these connective abnormalities are known to be risk factors for artery dissection or rupture [1]. Unfortunately, common imaging analyses (TC scan, MRI, Doppler sonography) could diagnose vascular clinical features only, while the microscopic abnormal distribution of collagen fibers forming the microstructure of the vascular wall underlies a silent arteriopathy known as “weakness of the vessel wall”, which is the real predisposing genetic risk for artery rupture and it does not present any clinical features or symptoms [2].

Ischemic and hemorrhagic vascular events, due to artery dissection or rupture, have been documented in these patients who usually claim headache, ipsilateral neck pain, incomplete Horner's syndrome (ptosis and myosis), bruits, transient ischemic attack, cerebrovascular accident signs such as amaurosis fugax,

syncope, encephalopathy and focal signs [2,3]. Cervicocephalic arterial dissections and rupture could be traumatic or spontaneous, and they could follow mild efforts as head and neck maneuvering, forceful cough and vomiting [4]. Few authors report a syndrome called “spontaneous cervical arterial dissection (SCAD)”, used to describe a nontraumatic tear or disruption in the wall of the brain-supplying arteries [5].

Here we report a female patient suffering from OI [6,7] who experienced a spontaneous rupture of internal carotid artery (ICA) during a neck dissection.

2. Case report

A 52-year-old female patient referred to our Institution for severe dysphonia and a left neck lump. Her medical history was positive for OI type I with several bone fractures and bleeding complications after adeno-tonsillectomy in 1980. The physical examination showed a right vocal fold paralysis, increased thyroid gland volume, and several bulky neck lymph nodes on both sides. One lymphadenopathy was 5 cm × 6 cm in size and seemed to be adherent to the right cervical neurovascular bundle at level II. A preoperative CAT scan of the neck and chest showed multiple calcifications in neck lymph nodes (from level II to level VI) bilaterally and a thyroid gland mass with similar calcifications

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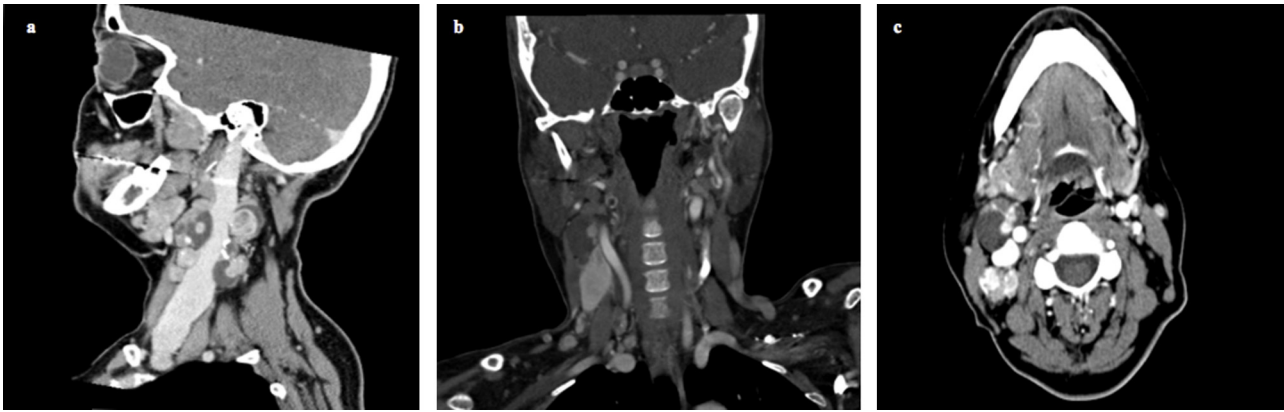


Fig. 1. TC scan imaging showing multiple cervical lymph node metastasis: (a) sagittal section, (b) coronal section and (c) axial section.

(Fig. 1). Vascular abnormalities were not evidenced. Subsequently, a fine needle aspiration biopsy was performed and a thyroid papillary carcinoma was found.

Thus, we performed total thyroidectomy with right recurrent nerve sacrifice and bilateral modified radical neck dissection including level VI.

During the right neck dissection, the cervical lymph node at sublevel IIA appeared to infiltrate the internal jugular vein (IJV) that was sacrificed (Fig. 2). During IJV ligation, manipulation of the vascular bundle determined a bleeding coming from a minimal leakage of ICA's wall which required its clamping. During these procedures we documented an evident retrograde blood flow from the Willis's Circle (Fig. 3). With the help of vascular surgeon, we removed the damaged tract of the ICA and he performed a transposition of the cranial stump of the ICA on the external carotid artery (ECA). The restoration of normal blood flow was intraoperatively confirmed by ultrasound echography. Finally, no cerebrovascular complications were found at the emergence from anesthesia.

The histopathologic evaluation confirmed a papillary carcinoma of the thyroid gland and bilateral neck metastases, pT4aN1b according to American Joint Committee on Cancer [8].

The histopathologic analysis of the damaged ICA tract did not show any cancer infiltrations of the artery's walls, while the signs of arterial wall dissection were documented (Fig. 4).

The patient was discharged 10 days later and she had not experienced any disease relapses after 12 months.

3. Discussion

Connective tissue disorders alter the structure of vascular walls, causing a silent arteriopathy known as “weakness of the vessel wall” [2], which does not present any clinical features and it is not diagnosed by common imaging techniques; dissection of carotid and vertebral arteries is reported as common feature in patients suffering from connective tissue disorders, but usually they claim alert symptoms. The most common locations of cervicocephalic artery anomalies are regions where these vessels are mobile, not firmly anchored to other arteries or bony structures. The ICA is relatively fixed proximally, at the origin from the common carotid artery, and distally, at the point of penetration in the petrous portion of the temporal bone. The arterial segment between these two points of anchorage is mobile and therefore vulnerable to stretching, especially during neck hyperextension with head rotation, which can stretch the ICA against an upper cervical vertebra or a prominent styloid process. The strain often occurs around 3 cm above the carotid bifurcation [5]. Usually, patients who experience cervical artery dissection or rupture might report either precipitating events or prior minor neck trauma [9], even if a clear cause–effect relationship is still debatable [10].

Our patient was affected by OI type I. Vascular complications are rare in OI when compared with other diseases involving connective tissue [6,7]. The most common vascular disorders are dysplasia of aortic and mitral valves, and congenital cardiac

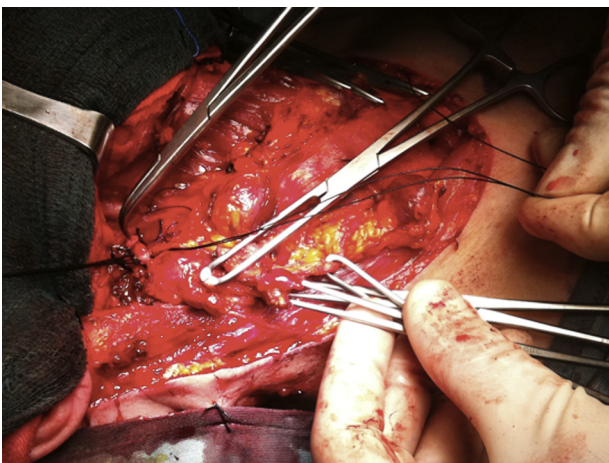


Fig. 2. Ligation of the right internal jugular vein (IJV) during neck dissection procedure: multiple and bulky lymph node metastases are visible in the surgical field. Two silky stitches have been placed around the cranial portion of the right IJV.

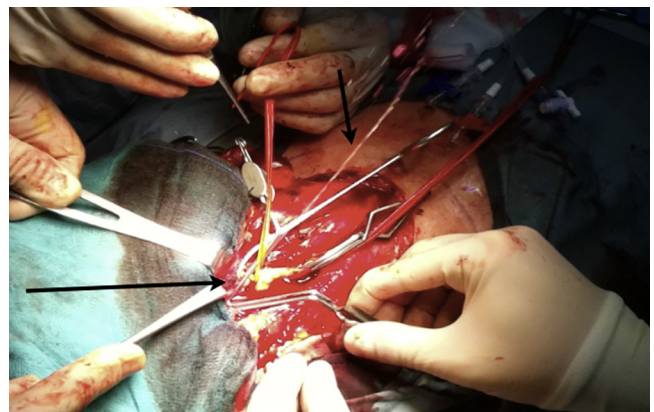


Fig. 3. Retrograde blood flow coming from the cranial resected stump of the internal carotid artery (ICA): arrows point the arterial blood jet coming from the cranial portion of the resected ICA. Red loops have been placed around the caudal stump of the ICA in order to perform the following anastomosis. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of the article.)

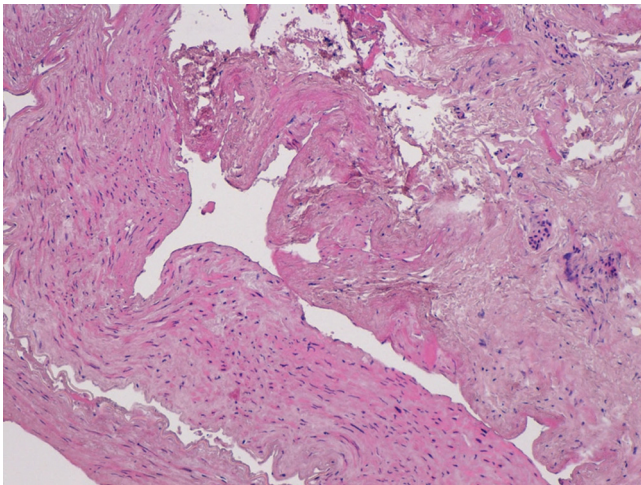


Fig. 4. ICA's wall alterations at optic microscopy: thickened intima and initial dissection at the adventitia level.

malformations. The ascending part of the aorta could be involved [11]. Spontaneous hemorrhages involving retina and conjunctivae, epistaxis and melena may occur due to capillary fragility. On the other hand, vascular cerebral complications are very rare [11]. Thus, the relationship between cervical artery events and OI is difficult to demonstrate [3].

Here we reported, to the best of our knowledge, the first case of spontaneous ICA's rupture that occurred during surgical neck dissection in a female patient suffering from OI with no evidence of vascular anomalies at the CAT scan. The rupture occurred during a gentle manipulation of this major vessel during a standard surgical procedure, without any direct trauma on it; thus, in our opinion, and in accordance with literature [9,10,12], the intrinsic wall weakness was the predisposing cause of its rupture. Moreover, the fact that it was not a catastrophic artery rupture steers toward considering the connective tissue disorder as the main risk factor for this kind of potential life-threatening event, excluding improper surgical approaches and the absence of reported neoplastic vessel infiltration.

In accordance with our experience, we assert that surgical manipulation of the carotid bundle, which can occur during neck dissection procedures, should be performed carefully in patients

with collagen disorders, because it could lead toward major vessel rupture and then to its sacrifice, with potential severe clinical consequences. Additional imaging techniques such as contrast-enhanced CAT scan, including angio-MRI, 3D-CTA, and carotid artery ultrasonography [1] might help in diagnosing the arteriopathy when evident, but the higher risk for spontaneous cervical artery rupture should be taken into consideration anyway.

Conflict of interest

All authors disclose no financial support or relationship that may pose a conflict of interest.

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