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Case Report

Acquired Phlebectasia of the Internal Jugular Vein in a Patient with Sjögren's Syndrome. An Unusual Accidental Finding

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Abstract

Internal jugular vein phlebectasia is a rare condition, presenting as a unilateral and usually painful neck swelling, mostly affecting the right side of the neck. The acquired subtype of internal jugular vein phlebectasia frequently affects adults and may be caused by trauma, thoracic outlet syndrome, tumors, local or systemic inflammatory conditions. Complementary tests are necessary to confirm the diagnosis. Imaging studies, such as eco-doppler, reveal the venous flow state and contrasted CT scans allow for the differentiation between solid tumors and vascular lesions. The purpose of this case report is to present an unusual case of an acquired internal jugular vein phlebectasia in a patient with Sjögren's Syndrome, presenting it as a chance finding and analyzing a possible mechanism to associate both entities.

Keywords: Vascular Malformations; Diagnostic Imaging

Abbreviations

IJVP: Internal Jugular Vein Phlebectasia; SS: Sjögren's Syndrome

Introduction

Internal jugular vein phlebectasia (IJVP) is a rare condition presenting as a unilateral and usually painful neck swelling, mostly affecting the right side of the neck [1]. This alteration is especially visible when the patient coughs or during a Valsalva maneuver [1]. Its etiology is uncertain, trauma and genetic factors are involved [1]. The acquired subtype of IJVP frequently affects adults and may be caused by trauma, thoracic outlet syndrome, tumors and local or systemic inflammatory conditions [2]. The purpose of this case report is to present an unusual case of IJVP in a patient with Sjögren's Syndrome (SS), presenting it as a chance finding and analyzing a possible mechanism to associate both entities. We could not find any study in the literature that refers to IJVP and SS, analyzing some possible mechanism to associate both diseases, as reported in our case.

Case Report

We present the case of a 66-year-old female patient, who attends presenting a painless increase in volume in the right parotid region of 4 days of evolution. Her medical record was relevant for recurrent episodes of acute sialadenitis, xerophthalmia, xerostomy

and xerovagina (episodes 2 - 3 times a year, without requiring hospitalizations for this reason, with a duration between 1 - 3 weeks, without identifying any predisposing factor). On physical examination, a firm swelling in the right parotid region was observed, with signs of inflammation (heat, blush, pain). The patient denies any history of trauma in the area. Intraorally, the right Stenon's duct presented scarce salivary flow without signs of purulent exudate. Differential diagnoses were considered, including exacerbated chronic sialadenitis and SS. A double intravenous antibiotic therapy with Ampicillin-Sulbactam and Clindamycin was started. Serologic blood tests were requested, and a positive SSA antibody was obtained, confirming the diagnosis of SS. A CT scan was also obtained where an enlarged right parotid gland with a rounded well-circumscribed isodense image at the level of the inferior pole was observed, congruent with an intraparotid abscess. As an incidental finding, a marked dilation of the right internal jugular vein was present (Figure 1), thus, an eco-doppler was performed showing discrete sclerosis of the vertebral carotid axes, absence of bulbar atheroma and a dilation of the right internal jugular vein without intrinsic or extrinsic pathology associated, confirming the diagnosis of IJVP (Figure 2). After 7 days of antibiotic treatment, there was a total remission of the inflammatory lesion in the right parotid gland. In a multidisciplinary discussion with the rheumatology and cardiovascular surgery services, a medical approach for SS and a conservative approach with periodic checkups for IJVP were decided.

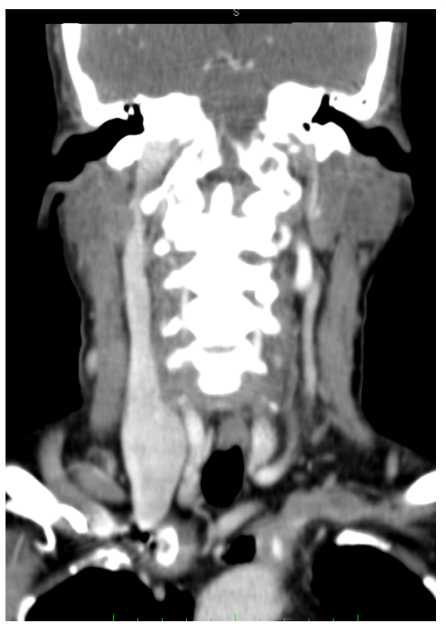

Figure 1: CT showing the dilation of the right internal jugular vein.

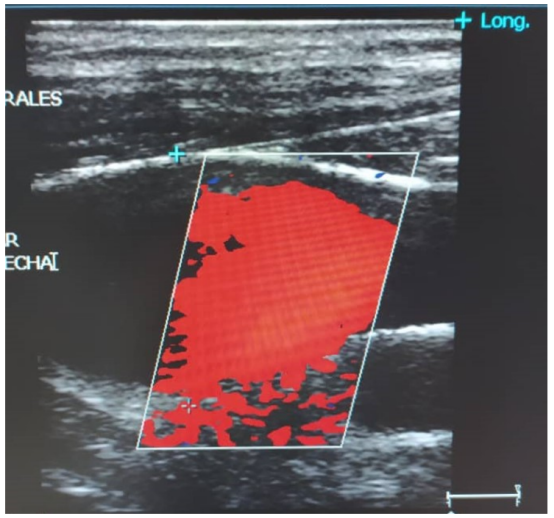

Figure 2: Eco-doppler performed during the valsalva maneuver showing dilation of the right internal jugular vein without intrinsic or extrinsic pathology associated.

Discussion

IJVP occurs due to a dilation of the jugular sinuses. This dilation has a multifactorial etiology: superior vena cava hypertension, anomalies in the vein walls, among others. Clinically, patients can refer pain, stiffness, cervical compression and dysphonia. A soft lateral cervical mass can be evidenced which can be present since infancy and its size can increase during Valsalva maneuvers [2,3]. In adult patients, this type of pathology is associated with acquired forms that can be caused by trauma, thoracic outlet syndrome, tumors and systemic inflammatory pathologies. In the case report presented, the etiology of the IJVP could be associated with a chronic systemic inflammatory disease such as SS [4]. Complementary tests are necessary to confirm the diagnosis. Imaging studies, such as eco-doppler during the Valsalva maneuver, reveal local dilatation of the internal jugular vein and venous flow state; contrasted CT scans allow for the differentiation between solid tu-

mors and vascular lesions, also demonstrate the dilatation of the IJVP [4]. Differential diagnoses include laryngocele, bronchial cyst, cystic hygroma, cavernous hemangioma, and superior mediastinal cysts. As presented in our case, most of the manifestations are asymptomatic, and its management is conservative. Imaging monitoring may be adequate [2].

Further studies are needed to corroborate an association between IJVP and SS; the latter, being considered a chronic systemic inflammatory disorder [5] and being this class of pathologies typified as etiological factors in the development of IJVP, could be included within the pathologies responsible for the development of the vascular affection. In the case presented in this study, the etiology of the IJVP could be associated with a chronic systemic inflammatory disease such as SS. As presented in our case, most of the manifestations are asymptomatic, and its management is conservative. Surgical treatment is indicated for symptomatic patients or cosmetic purposes.

Conclusion

IJVP should be considered as a differential diagnosis when a cervical mass is managed despite the unusual nature of the entity. During the review carried out for this investigation, no study was found in the medical literature that reflected the association between IJVP with SS as reported in our case. Further studies are needed to corroborate an association between IJVP and SS; the latter, being considered a chronic systemic inflammatory disorder, and being this class of pathologies typified as etiological factors in the development of IJVP, could be included within the pathologies responsible for the development of the vascular affection.

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Disclosures

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