Case Report / Olgu Sunumu

Cervical cystic vagal schwannoma mimicking a type 3 second branchial cleft cyst: a case report

Tip 3 ikinci brankiyal yarık kistini taklit eden servikal kistik vagal schwannoma: Olgu sunumu

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In this article, we report a rare case of a cervical cystic vagal schwannoma mimicking a type 3 second branchial cleft cyst clinically, radiologically and cytologically. Although schwannoma is rare, it should be considered in the differential diagnosis of cystic neck masses. This article suggests that Doppler ultrasonography, computed tomography and fine needle aspiration biopsy findings may not be sufficient to provide the correct preoperative diagnosis of cystic lateral neck masses and further imaging techniques may be required.

Key Words: Branchial cleft cyst; computed tomography; neck; ultrasonography; vagal schwannoma.

Bu yazıda, klinik, radyolojik ve sitolojik olarak tip 3 ikinci brankiyal yarık kistini taklit eden nadir bir servikal kistik vagal schwannom olgusu sunuldu. Schwannom nadir olmasına rağmen, kistik boyun kitlelerinin ayırıcı tanısında akılda tutulmalıdır. Bu yazı, kistik lateral boyun kitlelerinin ameliyat öncesi doğru tanısını koymak için Doppler ultrasonografi, bilgisayarlı tomografi ve ince iğne aspirasyon biyopsisi bulgularının yeterli olmayabileceğini ve daha ileri görüntüleme tekniklerinin gerekebileceğini önermektedir.

Anahtar Sözcükler: Brankiyal yarık kisti; bilgisayarlı tomografi; boyun; ultrasonografi; vagal schwannom.

Branchial cleft cysts (BCCs) are the most common congenital masses of the lateral neck and they usually become apparent in the second and third decades of life.[1-3] Second BCCs are the commonest branchial cleft anomalies, representing about 95% of cases.[4,5] They generally present as a slow-growing, fluctuant mass at the mandibular angle along the anterior border of the upper third of sternocleidomastoid.[1,3,4] The cystic lateral neck masses that can mimic BCCs include lymphangiomas, thyroglossal duct cysts, ectopic thymic cysts, dermoid and epidermoid cysts, necrotic adenopathies and cystic nerve sheath tumors.[2-4] It is not easy to differentiate BCCs from their mimics clinically and radiologically before surgery because they have similar

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properties such as being soft, slow-growing and painless cystic lesions.[2,4] Unlike BCCs, cervical vagal schwannomas are uncommon neurogenic tumors that originate from Schwann cells.[6] They usually occur between the third and fifth decades of life. Schwannomas of the head and neck region constitute up to 45% of all schwannomas.[6-8] Although schwannomas found in the neck are usually solid, cystic schwannomas are extremely rare in this area.[9] We report a case that appeared clinically, radiologically, and cytologically as a type 3 second BCC, but realized perioperatively to be a vagal cystic schwannoma. Histologic examination confirmed our diagnosis. We have to bear in mind the cystic schwannomas in the differential diagnosis of cystic neck masses and perform further radiological examinations to provide an accurate preoperative diagnosis.

CASE REPORT

A 41-year-old female presented with a one-year history of a painless, slow-growing mass at her right mandibular angle. She did not have any symptoms of infection on her neck mass. Otolaryngological examination revealed a 3 cm diameter mobile mass in the level 2 neck region. The remainder of the examination was unremarkable. Ultrasonography (USG) of the neck showed a well-circumscribed, septated, hypoechoic cystic mass, 3 cm in diameter (Figure 1). A Doppler USG showed a non-vascular lesion. Fine needle aspiration (FNA) of the mass was consistent with abscess content and no atypical cell was detected. Contrast-enhanced computed tomography (CT) scan of the neck demonstrated a well-defined, non-enhancing, hypodense lesion with a relatively high-density content, suggesting a complicated cystic lesion in the right side of the neck, extending from the angle of the mandible to the common carotid artery (CCA) bifurcation between the internal jugular vein (IJV) and the external and internal carotid arteries (ECA and ICA). This lesion was displacing the IJV posterolaterally and causing prominent narrowing in its lumen and displacing the ECA and ICA slightly medially. The lesion showed a small 'tail' extending between the ICA and ECA (Figure 2). In view of the clinical, cytological and radiographic findings, our preoperative diagnosis was a type 3 second BCC. Before surgery, informed consent was taken from the patient about the possible complications of the neck surgery. During surgery, it was seen that the lesion originated from the vagus nerve. The vagus nerve was strictly adherent to the tumor. Preservation of the vagus nerve was attempted but could not be achieved. The tumor was completely excised along with the vagus nerve engulfed by the tumor (Figure 3). Postoperatively, the patient had vocal cord palsy

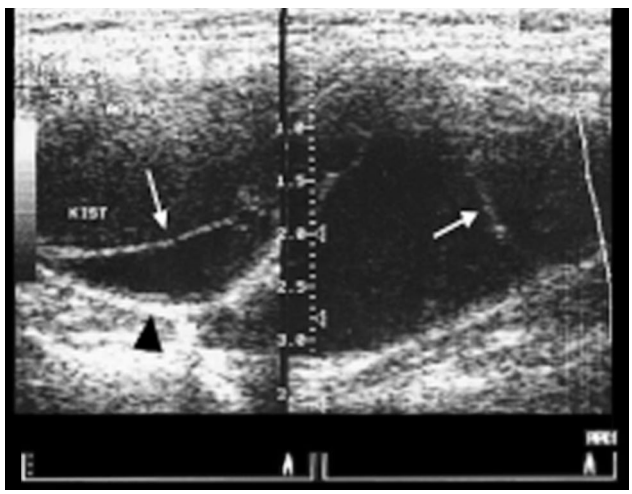

Figure 1. *Ultrasonographical images of the neck mass demonstrating a well-circumscribed (black arrow head points out the cyst wall), septated (white arrow), hypoechoic cystic lesion.*

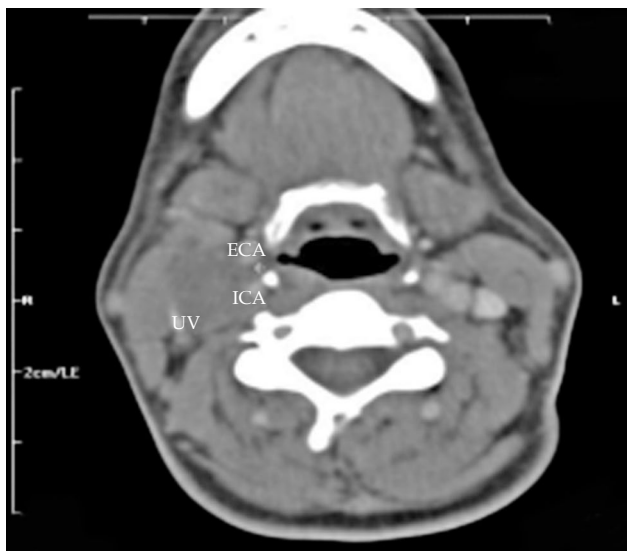

Figure 2. *Contrast-enhanced computed tomography scan of the neck, showing the high density cystic tumor extending between the internal (ICA) and external (ECA) carotid arteries and the internal jugular vein (IJV). The lesion showed a small 'tail' extending between the ICA and ECA (small arrow). The cystic tumor displaces the IJV posterolaterally causing prominent narrowing in its lumen.*

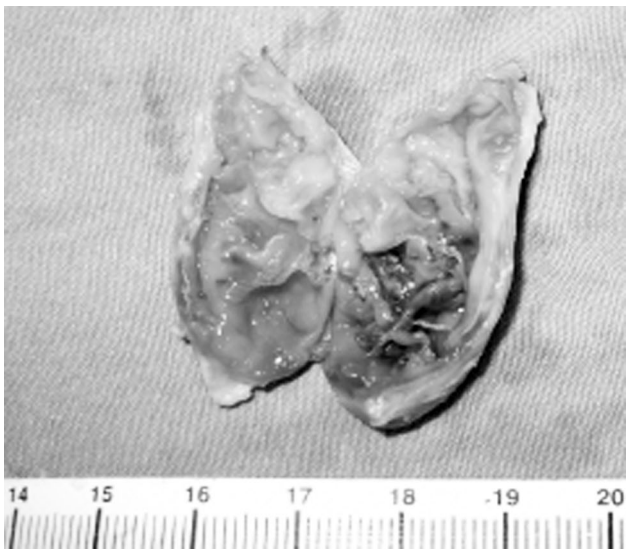

Figure 3. *Cystic vagal schwannoma divided into two pieces.*

on the operated side. Histologic examination of the specimen revealed a cystic schwannoma.

DISCUSSION

Branchial cleft cysts are the most common cysts that arise in the neck and second BCCs are the most common of the branchial anomalies.[2,4,5] Therefore a second BCC leads the list of diagnostic possibilities in younger patients with a painless, slow-growing, cystic neck mass at the mandibular angle.[4] Bailey classified second BCCs into four types based on their anatomic location.[4,10] Type 1 is beneath the platysma and at the anterior lateral surface of sternocleidomastoid muscle (SCM). Type 2 cysts are the most common and lie along the anterior surface of the SCM, just lateral to carotid space and posterior to the submandibular glands. Type 3 BCCs extend farther medially between the carotid bifurcation to the lateral pharyngeal wall. Computed tomography imaging may depict a tail sign of a type 3 cyst, extending between the ICA and ECAs.[2,10] Type 4 is the least common and lies within the submandibular and pharyngeal mucosal spaces, respectively.

Schwannomas are rare, benign, well-encapsulated tumors that may arise from any peripheral, cranial or autonomic nerve that has a schwann cell sheath.[6,11] Approximately 25 to 45% of extracranial schwannomas are located in the head and neck region and of these, a small percentage arise from the vagus nerve.[6,8,12] In the parapharyngeal space, schwannomas frequently arise from the vagus nerve and cervical sympathetic chain. Most of the tumors of this space are benign and solid in nature. The differential diagnosis for a lesion in this space includes salivary gland tumors, metastatic cervical nodes, BCCs, paragangliomas, lymphomas and neurofibromas. Vagal nerve and sympathetic trunk schwannomas made up 12% and 6% of the parapharyngeal space tumors in a series of 51 patients, respectively.[7] Kang et al.[8] described six patients with tumors of vagal origin from a total of 21 patients with head and neck schwannomas. Cervical vagal schwannomas are usually asymptomatic, solitary lesions which present as painless, slow-growing, lateral neck masses. Conservative surgical resection is the treatment of choice since they are benign lesions.

Cervical cystic vagal schwannomas are extremely rare. Preoperative diagnosis is difficult because they can mimic cystic lateral neck masses especially the most common second BCCs.[1,2] Due to their rarity, these tumors are generally not even taken into consideration in the differential diagnosis. Better imaging in the form of magnetic resonance (MR) imaging or CT scans and FNA cytological techniques are required for correct preoperative diagnosis.[1,2,8] In addition, Doppler USG may be useful in distinguishing cystic neck lesions. Gritzmann et al.[13] reviewed the usefulness of sonography in evaluating soft tissue masses of the neck. They reported that on color Doppler imaging, no vascularity is demonstrated inside the BCCs but schwannomas usually show moderate to significant vascularity. In our patient, Doppler USG showed a non-vascular lesion but it was not a BCC. Similar to our patient, Le Corroller et al.[14] reported a 43-year-old woman with cervical cystic vagal schwannoma whose color Doppler examination did not reveal internal vascularity. Therefore we can conclude that cystic schwannomas may not show vascularity on Doppler USG which is characteristic for schwannomas.

The utility of FNA biopsy in the preoperative diagnosis of schwannomas is limited. Colreavy et al.[6] reported that FNA gave a cytological diagnosis in two (25%) of eight head and neck schwannomas. Kang et al.[8] reported that FNA was performed on 10 patients with head and neck schwannomas and a definitive cytological

diagnosis of schwannoma was made only in two patients (20%), while in another three the diagnosis was suggested by the presence of spindle cells. In our case, the cytology was consistent with abscess content so it made us think of a cystic lesion.

On CT scans, schwannomas appear as well-defined, fusiform and intensely contrast-enhancing mass lesions. As the tumor enlarges internal cystic change becomes more prominent and it shows relatively homogenous contrast enhancement.[6,8] In our case, CT scans demonstrated a well-defined, hypodense, cystic lesion without contrast enhancement in the right side of the neck and depicted a tail sign of a type 3 second BCC, extending between the ICA and ECAs. This tail sign has been considered pathognomonic for type 3 BCCs.[2,4,10]

In this case, clinical, radiological, and cytological findings were considered sufficient to diagnose a type 3 second BCC. Saito et al.[7] reported that vagal schwannomas had been found to separate the carotid arteries from the IJV. They also reported that vagal schwannomas could displace the sheath vessels posteriorly, without splaying them. In our case, vagal schwannoma separated the carotid arteries from the IJV but extended between the ICA and ECA. Since the lesion was a pure cystic mass on Doppler USG and CT scans, we considered a second BCC first with no need for further investigation but before surgery, we could use other imaging techniques such as MR imaging, MR spectroscopy and MR diffusion-weighted imaging that may help differentiate schwannomas from second BCCs.[1,2]

In our review of the literature, this is the first reported case of a cervical vagal cystic schwannoma extending between the ICA and ECA mimicking a type 3 second BCC. Although schwannoma is rare, it should therefore be included in the differential diagnosis when a type 3 cyst is considered. Consequently, as in the case presented here, Doppler USG, CT and FNA biopsy findings may not be sufficient to provide the correct preoperative diagnosis of cystic lateral neck masses and further imaging techniques may be required.

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