Distant Adenocarcinoma Metastasis to Internal Jugular Vein From Unknown Origin Case Report

Bir Olguda İnternal Juguler Vene Odağı Bilinmeyen Uzak Adenokarsinom Metastazı

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Geliş Tarihi/*Received:* 11.06.2009
Kabul Tarihi/*Accepted:* 14.09.2009

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ABSTRACT Jugular vein distention as a neck mass is seen rarely and mostly the cause is tumoral invasion or intravascular trombosis. Tumoral invasions occur through metastatic lymph nodes or regional tumors. Distant neoplasms may cause also trombosis by creating coagulopathy or by pressure of lymphadenopathy. In this case, internal jugular vein was distended due to metastatic adenocarcinoma whose origin has not been found. There was not metastatic lymph node, regional tumor or intravascular trombosis. In English literature, no distant metastasis to jugular vein has been reported and from this aspect, this case may be the first one. As a conclusion, jugular vein occlusions must not be interpreted directly in favor of thrombus, even in the absence of regional primary tumor or metastasis, and in case of any doubts, further tests must be performed.

Key Words: Jugular veins; adenocarcinoma; neoplasm metastasis

ÖZET Boyunda kitle nedenleri arasında juguler ven distansiyonu nadir görülür ve çoğunlukla sebep direkt tümöral invazyon veya damar içi trombozdur. Tümöral invazyon, metastatik lenf nodu veya bölgesel tümör vasıtasıyla olur. Uzak neoplasmlar, aynı zamanda koagülopatiye yol açarak veya lenfadenopati basısıyla tromboza sebep olabilirler. Bu olguda orijini belli olmayan bir adenokarsinom metastazı sonucu internal juguler ven distansiyonu vardı. Ancak metastatik lenf nodu, bölgesel tümör veya damar içi tromboz yoktu. İngilizce literatür tarandığında juguler vene uzak metastazın rapor edilmediği görülmektedir, bu yönüyle olgumuz ilktir. Sonuç olarak bölgesel pimer bir tümör veya metastaz yokluğunda bile, her juguler ven oklüzyonu damar içi trombüs olarak değerlendirilmemeli, şüphe varsa daha ileri tetkiklere başvurulmalıdır.

Anahtar Kelimeler: Juguler toplar damarlar; adenokarsinom; tümör metastazı

Turkiye Klinikleri J Cardiovasc Sci 2010;22(1):128-31

Primary neoplasm originating from the internal jugular vein is very rare.[1] Metastasis of regional or distant tumors to cervical lymph nodes may be seen frequently, however invasion of carotid artery and/or jugular vein from lymph node metastasis is uncommon.[2,3] Jugular vein invasion by oral-maxillofacial and neck malignant tumors is possible, but not frequent.[4,5] Also distant neoplasms may cause trombosis by creating coagulopathy or by pressure of lymphadenopathy.

A case of 39-year-old male with metastatic adenocarcinoma in the left internal jugular vein without metastatic lymph node, regional tumor or trombosis is presented.

CASE REPORT

A 39-year-old previously healthy male patient admitted with painful left neck swelling for 2 months. It had been gradually growing in dimensions. Doppler ultrasonography (USG) was reported in favor of vein thrombosis, yet computerised tomography (CT) eliminated the thrombosis and was interpreted in favor of a mass (Figure 1). He was consulted with oncology department and finally it was agreed to remove the mass in order to abolish the pressure upon carotid artery and also to take a biopsy.

He was operated under general anesthesia, during the operation a 7 cm. segment of the jugular vein was found to be distended fusiformly, maximum diameter of the mass was approximately 2.5 cm (Figure 2). No lymph node was present. Internal jugular vein was removed with 2 cm. tumor-free segment at both edges, frozen section biopsy of the specimens taken from both edges did not determine any cancer cells. Then the edges were ligated.

The biopsy of the removed specimen reported an adenocarcinoma metastasis (mucinous carcinoma with neuroendocrine differantiation). Ultrasonography and positron emission tomography (PET) of whole body was carried out three times, however the primer origin of the metastasis could not be found. At the second year follow-up, the patient was healthy.

FIGURE 1: Longitudinal section of the neck in computerised tomography with contrast agent.

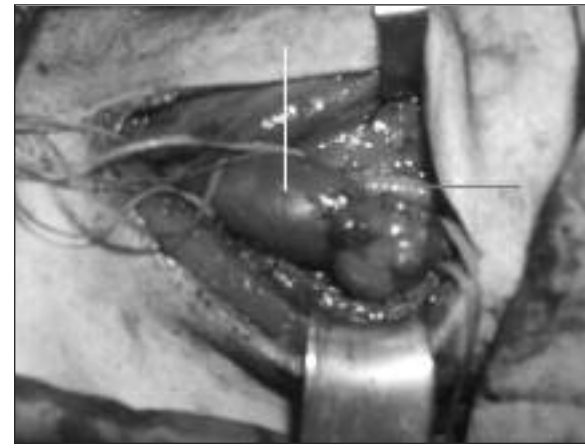

FIGURE 2: Enlarged jugular vein (yellow line) and common carotid artery (blue line) upward the vein (intraoperative view)

DISCUSSION

One of the uncommom causes of neck mass is jugular vein distension which is mostly related to trombosis of the vein due to pressure of regional lymph nodes and/or tumours. Trombosis of internal jugular vein was reported as a potentially serious pathology that can be spontaneous or secondary to head and neck infections, surgery, venous catheterization, intravenous drug abuse, thrombophilic states and also neoplasms most of which are regional.[6] Distant neoplasms may cause trombosis by pressure of metastatic lymphadenopathy or by creating coagulopathy (especially neuroendocrine tumors - Trousseau's syndrome).[7-9]

Incidence of deep venous thrombosis (DVT) involving the upper extremity accounts for approximately 4% of all DVTs. The most common predisposing factor is the presence of a central venous catheter, which is present up to 75% of patients with upper extremity DVT.[10] Marie et al.[11] reported that jugular vein thrombosis was 24.5% present in upper extremity DVT cases. In Marie et al.'s study, causative factors were malignancy (32.7%), venous catheters (22.4%) and thrombophilic states (8.2%).

In literature, there are some cases represented jugular vein invasion of metastatic lymph node or regional tumors. However, it is rare, because extranodal tumor spread must be present and mobility of the veins delay invasion.[2] Also in most of these cases, jugular vein trombosis develops. For unknown primary lesion, asymmetric enlargement of one or more cervical lymph nodes in an adult is almost always cancerous and usually is due to metastasis from a primary lesion in the mouth or pharynx. When adenocarcinoma is found in cervical lymph node biopsy, regional origins for metastasis are skin of ear, face, scalp, parotid, submandibular, salivary glands, thyroid gland. The most common sites for distant metastasis are pulmonary, esophageal, renal, ovarian, cervical and prostatic regions.

The exact incidence of cancer of unknown primary origin is not precisely known. It is almost certainly underreported, and its true incidence is most probably between 2-6% in the United States and 2-9% worldwide. In 15-25% of cases, the primary site can not be identified even on postmortem examination. In some cases, the original tumor may remain small or undetectable at the time of metastasis, leading to the clinical presentation of cancer of unknown primary origin. Whether a specific genetic or mutational factor plays a role in cancer of unknown primary origin remains uncertain. The inability to identify a primary site of cancer poses many challenges. The primary site of cancer usually dictates the treatment, expected outcome, and overall prognosis.

In this patient, during pre-operational examinations, Doppler USG has been reported in favor of vein thrombosis, yet CT has eliminated the thrombosis and was interpreted in favor of a mass. Also following surgical excision, no thrombus has been detected within the lumen and the lumen was completely obstructed with a mass. On this basis, it was concluded that CT was superior to Doppler USG in thrombus-mass distinction. This distinction was very important for approach to patient. In case it was thrombus, the patient would not be operated and anti-coagulant treatment would be initiated. However, when it is understood to be a tumoral mass, to prevent it from pressing on carotid artery and to make an open biopsy, I have resolved for surgery by obtaining the opinion of Oncology department.

Neither during surgery nor in PET (positron emission tomography) scans, performed during 20 months post-operative follow up, a mass or lymph node, which will cause direct invasion has not been detected. This was the interesting point. Because as far as I know, no distant metastasis to jugular vein has been reported and from this aspect, this case is a first. Although with pathological report, the presence of an adenocarcinoma is proven, the patient is administered chemotherapy, despite the fact that the failure to find the primary, made it impossible to apply surgery or radiotherapy, aimed to the primary. What is surprising and good is the fact that the health of the patient was excellent during this process and lack of any complaints in the surgical area or in any other location.

This case has shown that jugular vein distensions must not be interpreted directly in favor of thrombus, and in case of any doubts, Doppler USG must not be deemed sufficient and further tests must be performed and it must be considered that a remote metastasis to jugular vein may be in question without a regional mass.

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