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A case of angiosarcoma arising from internal jugular vein

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ABSTRACT

Primary angiosarcoma is a rare disease with a poor prognosis. It most commonly arises in the head and neck region; localization in the deep soft tissue of the neck is extremely rare. We herein present a case of angiosarcoma derived from the right internal jugular vein. A 79-year-old man presented with a 1-month history of a growing right neck mass. Computed tomography, magnetic resonance imaging, positron emission tomography-computed tomography, and fine-needle aspiration cytology revealed a malignant tumor of unknown origin. Right neck dissection was performed for both diagnosis and therapy. Immunostaining of the resected tumor cells revealed positivity for CD31, CD34, factor VIII-related antigen, and D2–40, which allowed for a definitive diagnosis of angiosarcoma. Postoperative radiotherapy (66 Gy) was performed on the right neck, including the surgical bed and upper mediastinum. The patient was followed up for 10 months with no recurrence. Only six cases of angiosarcoma arising in the deep soft tissue of the neck have been reported in the English-language literature. The present report is the first to describe angiosarcoma arising from the internal jugular vein.

1. Introduction

Primary angiosarcoma is a rare disease with a poor prognosis. Treatment is challenging in many cases, and the overall survival rate is about 35% [1]. Angiosarcoma can arise in any soft tissue structure or viscera [1]. The head and neck is the most common primary site (27.0% of all cases), followed by the extremities (15.3%), trunk (9.5%), liver, heart, bone, spleen, and other locations [1]. The scalp is the most common site for development of cutaneous angiosarcoma within the head and neck region. Development of angiosarcoma in the deep neck tissues, however, is extremely rare. We herein report an extremely rare case of angiosarcoma that occurred in the deep part of the neck.

2. Case report

A 79-year-old man presented with a 1-month history of a growing right neck mass. Physical examination revealed a soft, mobile, nontender mass that measured about 60 mm in diameter. No other lesions were apparent anywhere on his body, including the skin. Enhanced computed tomography showed a relatively well-defined and heterogeneous mass between the internal jugular vein and common carotid artery with a maximum diameter of 60 mm (Fig. 1). Magnetic resonance imaging also demonstrated a mass of heterogeneous density on both T1- and T2-weighted images.

Although fine-needle aspiration cytology of the mass suggested an adenocarcinoma, screening examinations such as thyroid ultrasound, laryngopharyngeal fiberscopy, upper gastrointestinal endoscopy, and colon fiberscopy showed no apparent primary lesions. The tumor was thought to be a cancer of unknown primary origin, and positron-emission tomography (PET) was performed to identify the primary tumor. Localized fluorodeoxyglucose accumulation (maximum standardized uptake value of 14.1) was seen within the right neck mass; however, no other abnormal accumulation was detected.

A right neck dissection was performed under a diagnosis of metastatic cervical carcinoma of unknown primary origin. The tumor was firmly adhered to the right internal jugular vein and sternocleidomastoid muscle; both of these structures were

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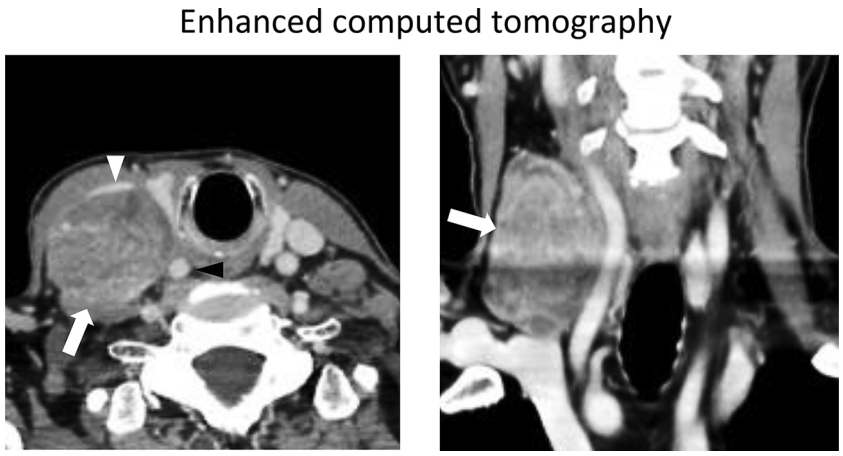

Fig. 1. An enhanced mass (arrow) with a diameter of 60 mm was found between the common carotid artery (black arrowhead) and internal jugular vein (white arrowhead). The internal jugular vein was displaced anteriorly by the mass. The mass contained several necrotic low-density areas and mildly enhanced solid areas. Invasion to surrounding areas was not apparent.

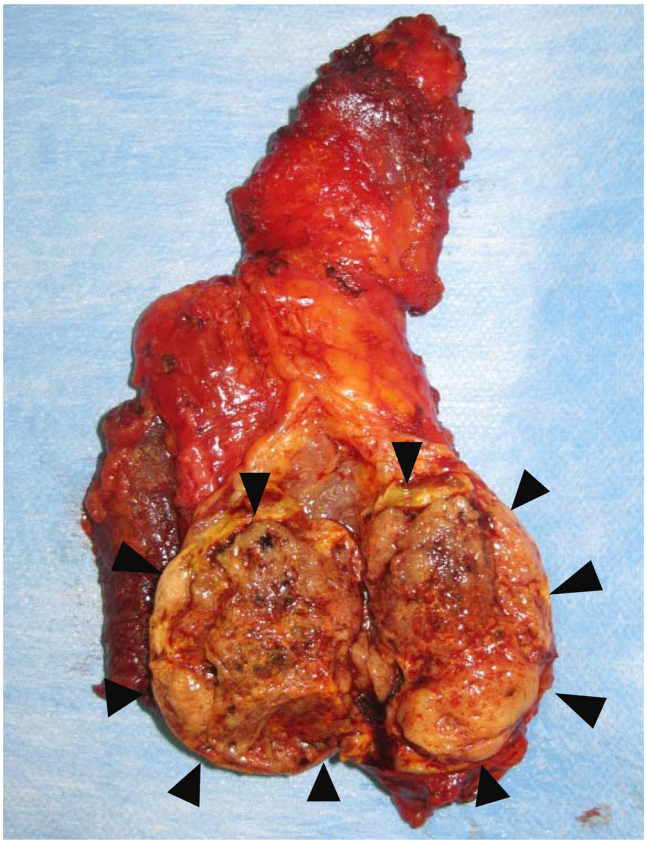

Fig. 2. The dissected specimen of the right neck. The mass contained yellow-brown solid components and multiple cystic components with dusky red fluid (arrowheads).

resected together. The common carotid artery and vagus nerve were not adhered to the lesion and were thus preserved. The largest diameter of tumor was 60 mm, and the cut surface showed yellow-brown solid components and multiple cystic components containing dusky red fluid (Fig. 2).

Hematoxylin–eosin staining of the excised specimens showed marked proliferation of spindle cells with irregularly shaped cell bodies and enlarged, irregular nuclei with prominent nucleoli. These findings suggested a cancer arising from both epithelial cells and soft tissues (Fig. 3A). The lumen of the internal jugular vein was patent and the tumor did not appear within it; however, the tumor was adjacent and fixed to the internal jugular vein. No lymph node tissue was seen within the tumor on histological examination. Immunostaining of the tumor cells revealed positivity for CD31, CD34, factor VIII-related antigen, and D2–40 and negativity for epithelial markers such as cytokeratin MNF116, CK7, CK20, and EMA. These pathological findings led to a diagnosis of angiosarcoma (Fig. 3B). The margin of the specimen and all excised lymph nodes were tumor-free. According to the American Joint Committee on Cancer and Union for International Cancer Control (AJCC/UICC) staging system for soft tissue sarcomas, the pathological stage of the tumor was III (T2bN0M0).

Postoperative radiotherapy (66 Gy) was performed on the right neck, including the surgical bed and upper mediastinum. The patient was followed up for 10 months with no recurrences.

3. Discussion

Angiosarcoma arising in the deep soft tissue of the neck is extremely rare; only six cases have been reported in the English-language literature (Table 1). Additionally, angiosarcomas rarely arise from major vessels. In the present case, the tumor was adjacent and fixed to the internal jugular vein on both surgical and pathological examinations, although there was no transformation zone from the tissue of the internal jugular vein. Thus, the tumor was most likely to have been an angiosarcoma derived from the right internal jugular vein. To the best of our knowledge, no other previous reports have described angiosarcoma arising from the internal jugular vein. It seems unlikely that the tumor was a metastatic lymph node because no apparent primary lesions on the patient's skin or other body parts were found on physical examination or PET. Moreover, the pathological examination showed no lymph node tissue within the tumor.

Four of the six previously reported cases of angiosarcoma arising from the deep soft tissue of the neck involved malignant transformation of a vagus nerve schwannoma to an angiosarcoma. Such angiosarcomas arise directly from the tumor vasculature as opposed to schwannoma cells [2].

The main histological feature of the angiosarcoma in the present case was dense growth of spindle cells with irregularly shaped cell bodies and irregular nuclei with prominent nucleoli. Vasoformation was not evident. An established histological pattern