

An ectopic hamartomatous thymoma compressing left jugular vein

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Abstract

Ectopic hamartomatous thymoma (EHT) is an extremely rare benign neoplasm. It is usually found at the root of the neck (frequently on the left) and does not usually impact adjacent tissues in clinically significant ways. While EHT manifests distinct pathological features, the lesion is either asymptomatic or may show nonspecific clinical features. We report one case of EHT which was assumed to be of low malignant potential since it severely compressed the inlet of left internal jugular vein as seen by computed tomography scan. To the best of our knowledge, this clinical finding of EHT is very rare. After the diagnosis and treatment of this patient, we believe that EHT or suspected EHT should be treated less invasively.

Key words: Ectopia, hamatoma, mini-sternotomy, thymoma

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Introduction

Ectopic hamartomatous thymoma (EHT) is a benign neoplasm that is usually located in the lower neck region.^[1] Surgical resection is often adequate for a cure because of its benign disposition.^[1,2] Several cases have been reported in the literature; however, most lack computed tomography (CT) imaging data.^[1-8] We report here one case of EHT where the left internal jugular vein was found to be severely compressed by the tumor when imaged by CT scan. To the best of our knowledge, this clinical manifestation of EHT is very rare. This discovery shows that not all cases of EHT are necessarily not impactive, and even so, the surgical approach should still be conservative.

Case Report

A 40-year-old male nonsmoker presented with a 2 week history of a painless neck mass that was slightly tender on palpation. There was no history of difficulty with swallowing, voice change, weight loss, fever, or intercurrent illness. On physical examination, a 5.0 × 3.0 cm, firm round mass was found to be associated with the posterior surface of

the head of the left clavicle. The mass was not attached to the adjacent soft tissue or thyroid gland, but it was firmly attached to the sternocleidomastoid. The patient's basic laboratory tests all showed results within normal ranges. CT scan was performed to evaluate the relationship between the mass and surrounding tissues [Figure 1] and revealed an oval mass in the left region of the neck. The mass was not enhanced with contrast. Both the left carotid artery and jugular vein were pushed to the rear owing to pressure from the soft tissue mass. The left jugular vein was severely compressed [Figure 1a] and the distal lumen was significantly dilated [Figure 1b].

We performed local excision through a limited upper sternotomy under general anesthesia. During the surgery, the tumor was found at the rear of the sternocleidomastoid. The tumor was well-circumscribed and solid with a complete envelope, and measured about 4 × 3 × 3 cm in size without continuity from the thymus or thyroid gland. The left internal jugular vein was also not affected. The lesion was well-margined from histology [Figure 2]

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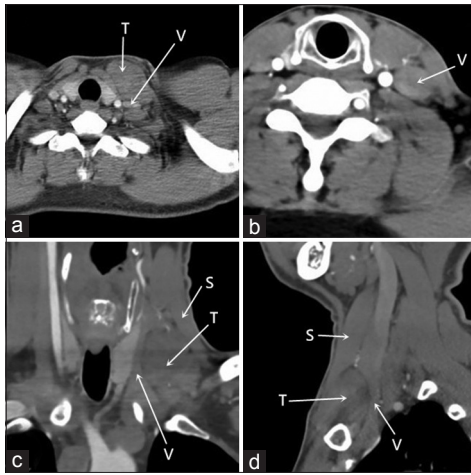


Figure 1: Computed tomography showing ectopic hamartomatous thymoma (“T” in figure) firmly attached to the sternocleidomastoid (“S” in figure) and the left internal jugular vein (“V” in figure)

and was composed of nests of epithelial cells, spindle cells, adipose cells, and a small amount of lymphocytes. Immunohistochemistry (IHC) showed that the lesion was diffusely positive for CD34, CK (Pan) (clone AE1/AE3), CK5/6, and CD10, but was negative for CK20, CD5, S100 protein, epithelial membrane antigen, and desmin. These findings were all consistent with the diagnosis of EHT.

The patient has remained disease free at 31 months.

Discussion

Ectopic hamartomatous thymoma was first described by Smith and McClure^[3] in 1982 and was named EHT by Rosai *et al.*^[4] in 1984. This tumor is found in a supraclavicular or retrosternal position and is located either superficial or deep to the sternocleidomastoid muscle in close proximity to the sternoclavicular joint.^[1] The incidence of this tumor is very low, with only dozens of cases reported in the literature.^[1-8] This tumor is pathologically characterized by a mixture of spindle cells, epithelial cells, mature adipose tissue, and lymphocytes.^[9] However, the histogenesis of EHT remains unclear. IHC study usually revealed myoepithelial differentiation of the spindle cells, suggesting that EHT is a mixed tumor composed of epithelial and myoepithelial cells. EHT may originate from the remnants of the cervical sinus of his, and may be more accurately named as branchial anlage mixed tumor.^[7]

Until date, classical EHT, which does not show evidence of cellular atypia, has never been reported to recur after complete excision. Since EHT follows a benign clinical pattern, conservative surgical excision is the first choice of treatment. Therefore, it is very important for clinicians to properly identify the tumor. A painless yet tender neck mass on palpation may be an initial sign of EHT. CT or magnetic resonance imaging (MRI) can be helpful in localizing the

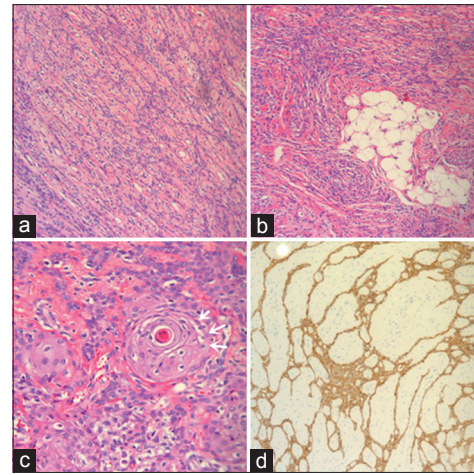


Figure 2: Histopathology and immunohistochemistry findings. (a) Epithelial cells with a reticulate arrangement H and E, ×100. (b) Spindle cells and adipose cells H and E, ×200. (c) Squamous differentiation and a squamous pearl (arrow) were identified in this lesion H and E, ×400. (d) Tumor cells are CK5/6 positive diaminobenzidine, ×100

tumor and verifying its attachments to other vital structures. Because of the mixed cellular components including epithelial cells, spindle cells and adipose cells, EHT usually appears as a well-circumscribed, heterogeneous mass, with mixed areas of fat and soft tissue density visible by CT or MRI.^[8,10] However, the present case is an atypical case, where the tumor appears to be a homogeneous mass. Unfortunately, most EHTs reported in the literature lack CT data, making it impossible for us to validate the usefulness of diagnosing EHT by CT. Practically, a diagnosis of EHT cannot be confirmed without postoperative pathology and IHC.

Tumors of the lower neck region present a significant challenge for the clinician. These tumors include conventional mixed tumors of skin adnexal or salivary gland origin, synovial sarcoma, a peripheral nerve sheath tumor variant, and cystic teratoma.^[7] Operating on several of these malignant tumors requires the surgeon to be highly experienced and skilled, and the main challenge is the ability to separate the tumor from surrounding blood vessels. Otherwise, the procedure may be limited to an incisional biopsy. In our case, our initial impression was that the lesion was a metastatic lymph node, that had been noted in previously reported cases.^[1,6] Since we considered the mass to be malignant, we decided to approach it through a limited upper sternotomy to achieve a wide access for vascular control. During the operation, we found the procedure to be easier than expected. Retrospectively, we believe that this tumor could have been removed through a simple cervical incision.^[2]

Our patient has maintained a good health since surgery. After his diagnosis and treatment, we now have more knowledge on EHT, which should be treated more conservatively than

other tumors of the lower neck region. However, it is still a challenge in accurately diagnosing EHT before surgery. Our experience is that CT may assist in diagnosing EHT and it is our hope that our report may assist future clinicians in their treatment of EHT or suspected EHT.

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