Cervical malignant teratoma masquerading as a hematoma: a case report

Han-Jie Lin[1], Chao-Yu Hsu[2,3] and
Stella Chin-Shaw Tsai[1,2] (iD)

Abstract

Background: Post-pubertal teratomas, which mostly occur at 20 to 40 years old, are more likely to be found at a metastatic site in up to 20% of cases and may be inadvertently overlooked. We present a case of cervical malignant teratoma that masqueraded as a hematoma.

Case presentation: A 24-year-old man presented to our institution with a 4-month history of a progressively relapsing painless mass in the neck, despite conservative treatments with oral medications. A huge space-occupying mass was identified with almost total occlusion of the left internal jugular vein. The likely diagnosis was an organized hematoma or congenital cystic tumor with internal hemorrhage. Surgical excisional biopsy of the mass lesion was conducted and a malignant teratoma was found. A whole-body positron emission tomography scan showed a left inguinal mass, bilateral intra-abdominal lymphadenopathies, and abdominal metastases. Histopathology further suggested the diagnosis of an immature testicular teratoma with multiple lymph node metastases. The patient received adjuvant chemotherapy with a bleomycin, etoposide, and cisplatin regimen. During follow-up, salvage second-line chemotherapy was required with a paclitaxel, ifosfamide, and cisplatin regimen.

Conclusion: Although uncommon, cervical teratoma should be taken into consideration once a painless and non-remitting mass lesion is found in a young adult.

Keywords

Cervical teratoma, neck metastasis, testicular neoplasm, chemotherapy, lymph node, germ cell tumor

[1]Department of Otolaryngology, Tungs' Taichung MetroHarbor Hospital, Taichung, Taiwan
[2]Department of Translational Medicine, National Chung Hsing University, Taichung, Taiwan
[3]Department of Urology, Tungs' Taichung MetroHarbor Hospital, Taichung, Taiwan

Corresponding author:
Stella Chin-Shaw Tsai, Tungs' Taichung MetroHarbor Hospital, 699 Section 8 Taiwan Boulevard, Wuqi, Taichung 434, Taiwan.
Email: tsaistella111@gmail.com

Introduction

Testicular cancer is the most common solid malignancy affecting young adults aged from 15 to 34 years old, even though it accounts for only 1% of all cancers in men.[1] Testicular teratoma is a germ cell tumor (GCT), and it comprises 95% of testicular cancers. Testicular teratoma may present in children as a prepubertal teratoma and in adult men as postpubertal teratoma. Prepubertal teratomas are mainly benign in nature and more differentiated, while postpubertal teratomas are usually a mixed variant neoplasm and are found at a metastatic site in up to 20% of cases.[2] The most common sites of metastasis of testicular teratoma are the lungs/mediastinum (49%), intra-abdominal lymph nodes (34%), cervical lymph nodes (18%), and liver (25%).[3] We present a case of cervical malignant teratoma that masqueraded as a hematoma.

Case report

A 24-year-old man presented to our institution with a 4-month history of a progressively relapsing painless mass in the neck, despite conservative treatments with antimicrobials and anti-inflammatory medications (Figure 1a). There was no associated dysphagia, shortness of breath, or hoarseness of voice. He had no prior surgery or irradiation to his neck. A clinical examination
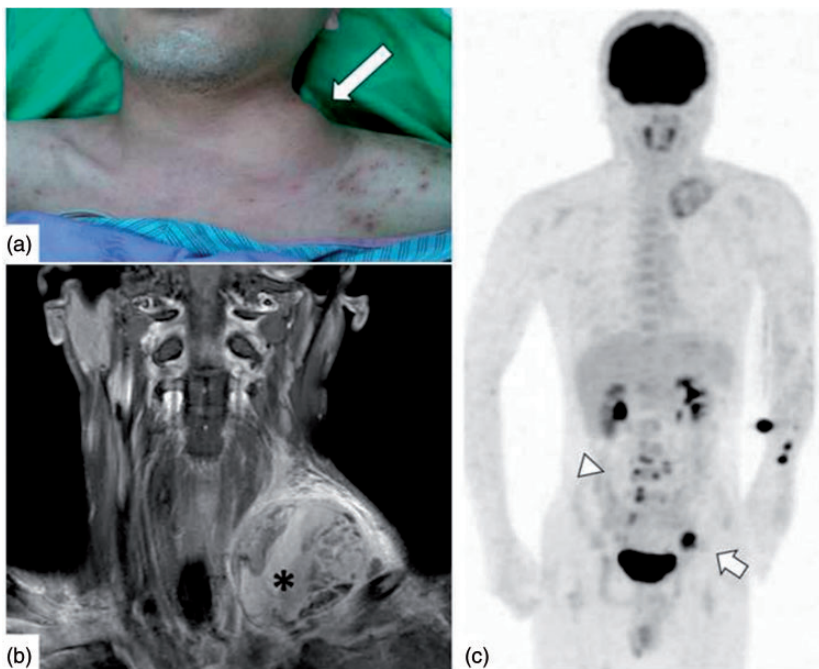

Figure 1. (a) Photograph of a large supraclavicular neck mass (long arrow) in the left side before incision of the skin. (b) Coronal view of a magnetic resonance imaging scan with contrast medium enhancement. A huge space-occupying neck mass of approximately 83 × 46 × 67 mm can be seen, with a heterogeneous short T1 signal without post-contrast enhancement at the site of the lesion and heterogeneous post-contrast enhancement at the lateral aspect (asterisk). (c) Coronal view of a whole-body positron emission tomography scan shows a left inguinal mass (short arrow) and bilateral intra-abdominal lymphadenopathies (arrowhead).

showed a firm, oval-shaped mass of approximately 7 cm in the long axis at level IV of the left neck, without local heat or pulsatile throbbing. Fiberoptic nasolaryngoscopy showed the nasopharynx without a mass lesion, a negative finding of a hypopharynx, and bilateral movable vocal cords. A head and neck contrast-enhanced computed tomography (CT) scan showed a huge space-occupying mass of approximately 51 mm with almost total occlusion of the left internal jugular vein. The mass contained heterogenous contents, which indicated a likely diagnosis of an organized hematoma or congenital cystic tumor with internal hemorrhage (Figure 2). Magnetic resonance imaging with a contrast study showed a huge space-occupying lesion (83 × 46×67 mm) at the left lower neck with a heterogeneous short T1 signal without post-contrast enhancement at the site of the lesion and heterogeneous post-contrast enhancement at the lateral aspect of the lesion. These findings also suggested the diagnosis of a complex tumor with internal hemorrhage or a pseudoaneurysm (Figure 1b).

A laboratory survey of tumor markers showed a large elevation in the α-fetoprotein level (459.2 ng/mL; normal range, <8.78 ng/mL) and the β-human chorionic gonadotropin level (449.2 mIU/mL; normal range, <5 mIU/mL). However, levels of carcinoembryonic antigen (3.1 ng/mL; normal range, <5 ng/mL), squamous cell carcinoma antigen (1.7 ng/mL; normal range, <1.5 ng/mL), lactate dehydrogenase
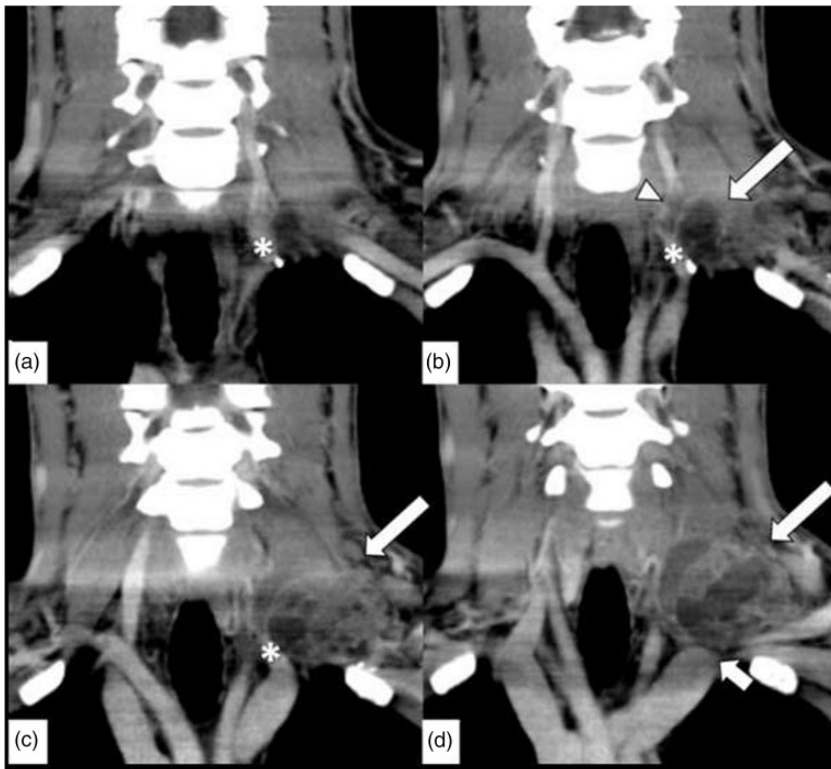

Figure 2. Image sequences of a contrast-enhanced computed tomography scan (from a to d) showing a mass lesion (long arrow) situated at the junction of the left internal jugular (asterisk) and subclavian veins (short arrow), with almost total occlusion of the left internal jugular vein (arrowhead).

(133 U/L; normal range, 106–221 U/L), and amylase (84 U/L; normal range, 20–112 U/L) were within the normal range.

Surgical extirpation was uneventful and showed a large tumor at the left lower neck in the carotid space compressing the internal jugular vein and it was 10 cm in length. The left subclavian artery and vein were ruptured during resection, and the thoracic duct inlet was exposed. All three structures were meticulously repaired. Postoperatively, the patient was well without a hematoma, surgical wound infection, or subsequent long- and short-term consequences. The entire cervical lesion measured $7.4 \times 6.8 \times 4.3$ cm and it was sent for a pathological examination. This examination showed primitive mesenchymal, ectodermal, and endodermal elements, including prominent primitive neuroepithelial cells, immature cartilage (Figure 3a), a primitive gland, and respiratory type epithelium (Figure 3b), consistent with immature teratoma of high grade. A whole-body positron emission tomography scan was performed and further showed a left inguinal mass (Figure 1c). Other findings were lesions in bilateral intra-abdominal lymphadenopathies and there appeared to be abdominal metastases. Scrotal ultrasonography also showed a 2.0-cm hypoechoic

mass and a 1.8-cm cystic mass mixed with a hypoechoic mass in the left testicle. The patient received another left radical inguinal orchiectomy and left inguinal lymph node dissection. A 2.5-cm palpable firm tumor within the left testis was excised with a 5-cm lymphadenopathy at the left internal ring of the inguinal canal.

A final histopathological examination of immunohistochemical staining showed the following: α-fetoprotein, focal positive; CD30, focal positive; β-human chorionic gonadotropin, focal positive; CD117, negative; and cytokeratin, focal positive. These findings confirmed the diagnosis of left testicular immature teratoma of high grade, without extending through the tunica albuginea, and with positive left inguinal lymph node metastases. Adjuvant chemotherapy with a bleomycin, etoposide, and cisplatin regimen was administered soon after completion of surgical interventions. Persistent elevated β-human chorionic gonadotropin levels were observed after a full course of four cycles of monthly chemotherapy with an episode of flare-up (from 7.55 to 34.44 mIU/mL), which suggested relapse. Salvage second-line chemotherapy was initiated with a paclitaxel, ifosfamide, and cisplatin regimen thereafter.
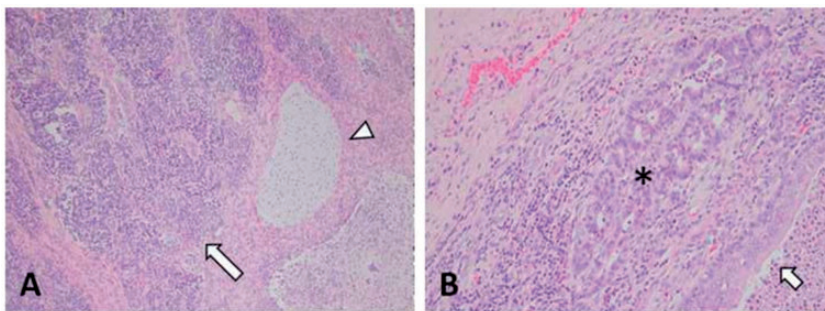

Figure 3. Histopathology shows that the tumor is composed of primitive elements of mesenchymal, ectodermal, and endodermal elements. (a) Prominent primitive neuroepithelial cells (long arrow) and immature cartilage (arrowhead) can be seen (hematoxylin–eosin staining, 40×). (b) High-power section showing a primitive gland (asterisk) and respiratory-type epithelium (short arrow) (hematoxylin–eosin staining, 100×).

Discussion

The initial suspected cause of the painless neck mass in the supraclavicular fossa in our patient was considered to be a hematoma due to rupture from adjacent vessels with suspected internal hemorrhage. We also suspected an arteriovenous fistula after performing magnetic resonance imaging because of the mass's painless, noninflammatory, non-pulsatile presentation. Other possible causes of the mass included the following: noninfectious inflammatory disorders, such as Castleman disease, sarcoidosis, Kikuchi–Fujimoto disease, Rosai–Dorfman disease, and other autoimmune diseases; infectious causes, such as bacterial lymphadenopathy, reactive viral lymphadenopathy, and tuberculous lymphadenitis, especially the population in Taiwan; congenital causes, such as branchial cleft cyst, cystic lymphangioma (previously known as cystic hygroma), complex cyst, ectopic thyroid gland, dermoid cyst, thymic cyst, teratoma, and paraganglioma; and malignancies, either primary (e.g., lymphoma, sarcoma, and paraganglioma) or metastatic (e.g., from the aerodigestive tract or thyroid, or infra-diaphragmatic genitourinary cancer).[4] However, after watchful waiting, the mass lesion did not show any sign of resolution. A serological examination did not show any major findings. A biopsy of the lesion was generally recommended for further evaluation. Because of hemorrhagic potential during the procedure, fine needle aspiration for a cytological study was not planned. The tumor was considered resectable en bloc. Therefore, the patient underwent surgical intervention, whereupon the final diagnosis was then established.

Saylor et al.[5] reported a case of a 44-year-old man with neck pain and swelling on the left side of the neck after uncomplicated placement of an implantable cardioverter-defibrillator through the left cephalic vein 12 weeks before presentation. Ultrasonography showed a heterogeneously hypoechoic lesion in the left supraclavicular region, with a size of 3.9 cm. A contrast-enhanced CT scan showed a lesion buried deep in the sternocleidomastoid muscle, and it abutted the left common carotid artery and brachial plexus. Needle aspiration of the mass revealed thick red fluid, which was consistent with blood. The mass initially shrank after the procedure, but recurred after 2 weeks. Trans-brachial arteriography was performed to evaluate possible injury to the subclavian vessels. The results of this examination were normal without extravasation, but a vascular blush was observed in the neighboring tissue. A biopsy was carried out at a later date and a pathological examination led to a diagnosis of mature teratoma. After reviewing the patient's history of orchiectomy for testicular cancer at 20 years old, a mixed GCT with a component of immature teratoma was diagnosed. Saylor et al.[5] concluded that metastatic immature teratoma with maturation of mesenchymal components was the most likely diagnosis.

GCTs of the testis are a common malignancy in men aged 20 to 40 years.[1] Testicular teratomas are nonseminomatous GCTs, which among others, also include embryonal cell carcinoma, yolk sac tumor, and choriocarcinoma. GCTs are derived from abnormal development of pluripotent stem cells, and consist of ectodermal, mesodermal, and endodermal germ cells with different degrees of maturation and differentiation. All GCTs are further subdivided into gonadal GCTs and extra-gonadal GCTs, and 2% to 10% of GCTs are attributed to extra-gonadal GCTs.[6] Teratomas have been found in various sites and organs. Gonadal teratomas occur in the testes or ovaries, or as extra-gonadal teratomas originating most commonly from midline structures, such as the retroperitoneal area, anterior mediastinum, or

sacrococcygeal region in decreasing order of the incidence rate.[7]

Based on findings of genetic studies, teratomas are considered as a single entity, regardless of the degree of maturation and differentiation.[3] Nonseminomatous histologies of teratomas, including embryonal cell carcinoma, yolk sac tumor, choriocarcinoma, and teratoma, account for approximately 50% of all GCTs. Only 2% to 6% of nonseminomatous GCTs are composed of a pure teratoma, which is not associated with serum tumor markers. Therefore, teratoma is frequently present in combination with other histological subtypes. Consequently, the name of a mixed germ cell tumor is suggested and frequently encountered when there is elevation of serum tumor markers, representing coexistence of other nonseminomatous GCT components.[8] Microscopically, mature elements contain three well-differentiated germ cell elements resembling normal postnatal tissue and they are thought to be benign without metastatic behavior. However, immature elements contain specific immature neuroepithelial tissue resembling that of the undifferentiated embryonic neural tube, and they were formerly called malignant teratoma. However, regardless of grading of the degree of immaturity in such tumors in adults, teratomatous GCTs can be found with similar characteristics to invasive malignant GCTs. Additionally, teratomatous GCTs may have already metastasized at presentation, even if they appear totally mature.[5]

Head and neck presentation of metastatic GCTs is rare, with neck metastases reported in 4% to 5% of all patients with testicular cancer.[9,10] Metastatic testicular GCTs typically initially involve the retroperitoneal lymph nodes and supradiaphragmatic mediastinal lymph nodes. These tumors then spread via the thoracic duct to its emptying site near the junction of the left internal jugular and subclavian veins.[11] Therefore, the left supraclavicular region is a plausible location where testicular teratomas may metastasize. Because testicular carcinoma is the most common malignancy in men aged 20 to 30 years, a left supraclavicular mass in this age group should raise suspicion for a concomitant testicular mass.[12]

Mehra et al.[13] conducted a retrospective review and found that 74% of patients had a neck mass at the initial diagnosis, and 67% of those had a neck mass as the presenting symptom, leading to a diagnosis of GCTs. In their study, preoperative tumor markers were also obtained and were normal in 58% of the patients, and were not significantly correlated with positive neck pathology, disease-specific survival, or overall survival. However, another study showed that persistently elevated tumor markers after first-line chemotherapy indicated a worse outcome for patients with metastatic GCT.[14] This finding is similar to our patient's serological values after the first adjuvant chemotherapy.

Conclusion

We experienced a rare, but insidious, cause of neck swelling in a young patient. Our findings suggest that extraordinary and potentially lethal causes of neck swelling should be taken into account in similar clinical situations. Although uncommon, cervical teratoma should be taken into consideration if it presents in a young adult as a painless and non-remitting mass lesion. A thorough review of systems and whole-body physical examinations are considered more important than a preliminary imaging study, especially when an origin other than the head and neck is suspected for a mass.

Ethics statement

This study was approved by the institutional review board of Tungs' Taichung MetroHarbor Hospital, Taichung, Taiwan (IRB#: 108075). Informed written consent was obtained from

the patient for publication of this case report and accompanying images.

ORCID iD

Stella Chin-Shaw Tsai 🄳 https: //orcid.org/0000-0003-2694-1207

References

1. Siegel RL, Miller KD and Jemal A. Cancer statistics, 2019. *CA Cancer J Clin* 2019; 69: 7–34. https: //doi.org/10.3322/caac.21551.
2. Wetherell D, Weerakoon M, Williams D, et al. Mature and immature teratoma: a review of pathological characteristics and treatment options. *Med Surg Urol* 2014; 3: 124.
3. Montironi R, Cheng L, Scarpelli M, et al. Pathology and genetics: tumours of the urinary system and male genital system: clinical implications of the 4th Edition of the WHO classification and beyond. *Eur Urol* 2016; 70: 120–123.
4. Flint PW and Thomas JR. Cummings otolaryngology. In: Flint PW and Cummings CW (eds) *Neoplasms of the neck*. 6th ed. Maarssen, Netherlands: Elsevier Gezondheidszorg, 2015, pp. 1787–1804.
5. Saylor PJ, Dudzinski DM, Mendoza DP, et al. Case 14-2019: a 44-year-old man with neck pain and swelling. *N Engl J Med* 2019; 380: 1854–1863.
6. Shirkhoda M and Soltani E. Management of cervical lymph nodes metastasis of nonseminomatous extra gonadal carcinoma - report of a rare case. *Clin Oncol* 2018; 3: 1540.
7. Carver BS, Al-Ahmadie H and Sheinfeld J. Adult and pediatric testicular teratoma. *Urol Clin North Am* 2007; 34: 245–251.
8. Milose JC, Filson CP, Weizer AZ, et al. Role of biochemical markers in testicular cancer: diagnosis, staging, and surveillance. *Open access J Urol* 2011; 4: 1–8.
9. Van Vledder MG, Van Der Hage JA, Kirkels WJ, et al. Cervical lymph node dissection for metastatic testicular cancer. *Ann Surg Oncol* 2010; 17: 1682–1687.
10. See WA, Laurenzo JF, Dreicer R, et al. Incidence and management of testicular carcinoma metastatic to the neck. *J Urol* 1996; 155: 590–592.
11. Dearnaley D, Huddart R and Horwich A. Regular review: managing testicular cancer. *BMJ* 2001; 322: 1583–1588.
12. Gavriel H and Kleid S. Benign neck metastasis of a testicular germ cell tumor. *Int Surg* 2015; 100: 164–168.
13. Mehra S, Liu J, Gupta A, et al. Cervical metastasis of germ cell tumors: evaluation, management, complications, and outcomes. *Laryngoscope* 2012; 122: 286–290.
14. Weisberger EC and McBride LC. Modified neck dissection for metastatic nonseminomatous testicular carcinoma. *Laryngoscope* 1999; 109: 1241–1244.