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Case Report

An Unusual Presentation of Growing Teratoma Syndrome

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Abstract

The growing teratoma syndroma (GTS) was first defined in 1982 as growth in masses despite the decrease of serum tumor markers after treatment with cytotoxic chemotherapy in non-seminamatous germ cell tumors. Due to the rarity of the GTS, tumor growth should not be mistaken for disease progression. Because this may cause oncological overtreatment in some patients. GTS is most common in the retroperitoneal region. However, it has been reported elsewhere (lung, lymph nodes, mediastinum, liver, and pineal gland). Since GTS is not responsive to chemotherapy and radiotherapy, the gold standart of treatment is the radical surgical excision of the mass. There are very few cases of GTS in the literature that develop from the supraclavicular lymph node. Here we present an unusual case of GTS at left supraclavicular and subcarinal lymph nodes.

Keywords: Growing teratoma syndrome, germ cell tumor, testis tumor.

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The growing teratoma syndroma (GTS) was first defined in 1982 as growth in masses despite the decrease of serum tumor markers after treatment with cytotoxic chemotherapy in non-seminamatous germ cell tumors (NSGCT).[1] Germ cell cancers are the most common solid tumors among men aged 15-40 years and it accounts for 1% of the male tumors.[2] Since GTS is a rare entity, it can be mistakenly evaluated as disease progression. Growing teratoma syndroma usually occurs in the retroperitoneum.[3] Surgical treatment is the only option for cure, since teratomas do not respond to chemotherapy and radiotherapy. Here we present an unusual case of GTS at left supraclavicular and subcarinal lymph nodes.

Case Report

We present the case of a 29 year old man who had surgery in 2010 due to undescended left testicle. There was no malign pathology report and any treatment history. In April 2018, the patient admitted to the hospital because of palpabl mass in the right testicle. Before surgery alpha fetoprotein (AFP) level was detected as 9,2 ng/mL (normal range 0-5 ng/ml), B-HCG was within normal levels and right partial orchiectomy was performed. Histopathological examination described as mixt germ cell tumor (seminoma %10, embryonal carcinoma %40, yolk sac tumor %10 and teratoma %40), pT2. Tumor was limited to testicular tissue and all surgical margins were intact. But, in-situ neoplasm continues at the parenchymal surgical margin. Abdominal tomography (CT) evaluation showed a few lymph nodes (measuring 7 mm) in the left paraaortic region. Post-operative AFP level was within normal levels (1,6 ng/mL). The patient did not accept total orchiectomy and radiotherapy was given to the rigth residual testicle (2000 cGy). 2 cycles BEP (Bleomycin, etoposide, cisplatin) was planned after radiotherapy but patient did not accept to receive chemotherapy. After 8 months, in November 2018, he admitted with palpable left supraclavicular lymphadenopathy. AFP was detected as 8,4 ng/mL, B-HCG was within normal levels. Thorax and abdominal to-

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mography evaluation showed no abnormal finding in addition to supraclavicular area. Biopsy was performed from left supraclavicular lymph adenopathy. Pathological evaluation reported as metastasis of testicular origin malignancy. The patient was treated with 3 cycles of bleomisin-etoposide-cisplatin (BEP) combination. Despite 3 cycles BEP treatment, post-treatment tomography showed enlargement of the left supraclavicular lymphadenopathy (from 36x27 to 45x28 mm). Re-biopsy was performed and the pathology resulted as a cyst. AFP was undetectable and the patient was planned to follow in a multidisiplinary tumor council. After 4 months of completion treatment left supraclavicular lymph node enlargement lymphadenopathy was continued to grow (from 45x28 mm to 61x47 mm) and newly developed subcarinal lymphadenopathy (24 mm) was detected. The patient's AFP was still undetectable. Because of the radiologically similar lymphadenopathies, GTS was considered in the foreground. Left supraclavicular lymph node excision was performed (Figs. 1, 2). Pathology was a reported as teratoma. Subcarinal lymph node excision was then performed (Fig. 3). Its pathology result was also reported as a teratoma. The patient is still being followed in remission.

Discussion

GTS is a uncommon clinical condition in patients with germ cell tumors. Most of the diagnosed cases are often in men, because germ cell cancer is more prevalent in men. The prevalence of GTS in metastatic nonseminomatous germ cell tumor is between 2.0% - 7.6% however it may rarely originate from pure seminoma.[4]

There is no definitive information about the etiology of GTSs. However, according to the most accepted opinions: The first may be that chemotherapy destroys immature malignant cells and the growth of mature teratomatosis components are not affected with treatment, and the other may be chemotherapy changing cell kinetics and convert some of the malignant cells to benign teratoma transformation.[3]

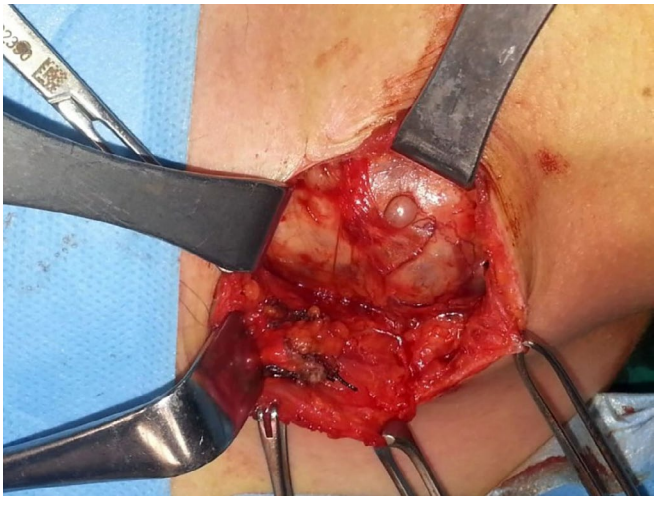

Figure 1. Supraclavicular lymph node excision.

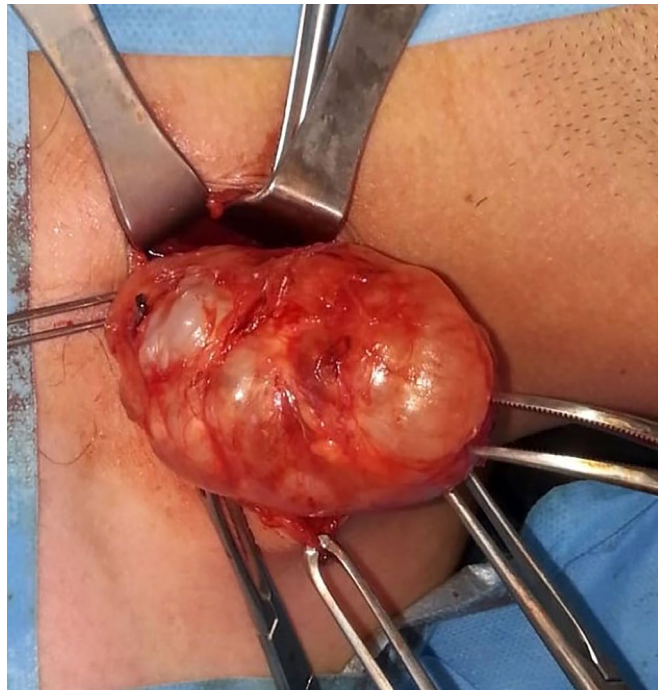

Figure 2. Supraclavicular lymph node excision.

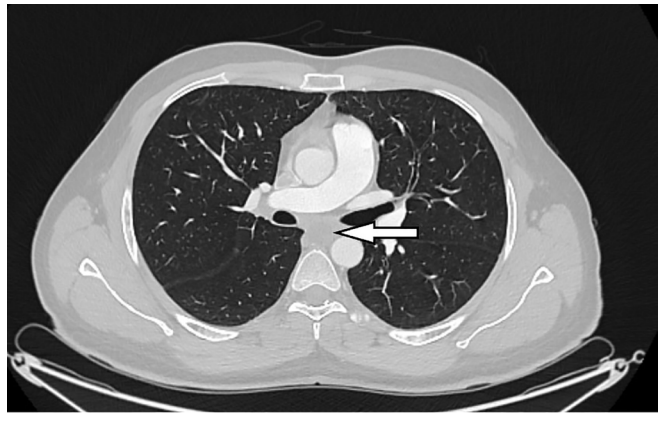

Figure 3. Subcarinal lymph node.

It is well-known fact that growing teratomas are benign, but they can grow by 0.5–0.7 cm per month, and the volume increase can be between 9.2-12.9 cm^3 per month,[5, 6] of course, growth trends may vary. It is also difficult to predict the behavior of GTSs in terms of local spread and malignant degeneration.

According to Andr´e et al., there are some clues to suggest GTS development: the presence of mature teratoma in the primary NSGCT, no reduction in tumor size during che-

motherapy; and teratoma in postchemotherapy residual masses.[4] Following closely the patient during and after chemotherapy is important. The first sign of GTS may be tumor growth during or after chemotherapy. Usually tumor growth is not accompanied by tumor marker elevation. However, rarely, small increases in tumor markers have been reported.[7] The presence of mature teratoma and absence of viable germ cells in the pathology is another finding that supports GTS.

Since GTS is not responsive to chemotherapy and radiotherapy, the gold standart of treatment is the radical surgical excision of the mass. Delayed surgery can cause local complications (postrenal failure or bowel obstruction, bile duct, or large vessel compression etc.). Therefore, it is recommended to perform surgery as soon as possible. Patients with GTS have heterogeneous clinical outcomes. During follow-up, 4-17% of patients relapse despite surgical complete resection.[8] Median recurrence time was reported as 48 months in one study.[7] In another study, 2 of 22 GTS patients developed recurrence at 2 and 6 months.[9]

GTS is most common in the retroperitoneal region. However, it has been reported elsewhere (lung, lymph nodes, mediastinum, liver, and pineal gland).[10, 11] There are very few cases of GTS in the literature that develop from the supraclavicular lymph node.[10, 12] Therefore, we think that the case of GTS, which rarely occurs in the supraclavicular and subcarinal lymph nodes, is special.

In conclusion, due to the rarity of the GTS, tumor growth should not be mistaken for disease progression. This may cause oncological overtreatment in some patients. Therefore, in the case of tumor growth with normal tumor markers, as in our case, considering GTS is the first and most important part of the treatment.

Disclosures

Informed Consent: Written informed consent was obtained from the parents of the patient for the publication of the case report and the accompanying images.

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References

1. Logothetis CJ, Samuels ML, Trindade A, Johnson DE. The growing teratoma syndrome. Cancer 1982;50:1629–35.
2. Shanmugalingam T, Soultati A, Chowdhury S, Rudman S, Van Hemelrijck M. Global incidence and outcome of testicular cancer. Clin Epidemiol 2013;5:417–27.
3. Gorbatiy V, Spiess PE, Pisters LL. The growing teratoma syndrome: Current review of the literature. Indian J Urol 2009;25:186–9.
4. André F, Fizazi K, Culine S, Droz J, Taupin P, Lhommé C, et al. The growing teratoma syndrome: results of therapy and long-term follow-up of 33 patients. Eur J Cancer 2000;36:1389–94.
5. Spiess PE, Kassouf W, Brown GA, Kamat AM, Liu P, Gomez JA, et al. Surgical management of growing teratoma syndrome: the M. D. Anderson cancer center experience. J Urol 2007;177:1330–4; discussion 1334.
6. Lee DJ, Djaladat H, Tadros NN, Movassaghi M, Tejura T, Duddalwar V, et al. Growing teratoma syndrome: clinical and radiographic characteristics. Int J Urol 2014;21:905–8.
7. Jeffery GM, Theaker JM, Lee AH, Blaquiere RM, Smart CJ, Mead GM. The growing teratoma syndrome. Br J Urol 1991;67:195–202.
8. Carver BS, Serio AM, Bajorin D, Motzer RJ, Stasi J, Bosl GJ, et al. Improved clinical outcome in recent years for men with metastatic nonseminomatous germ cell tumors. J Clin Oncol 2007;25:5603–8.
9. Paffenholz P, Pfister D, Matveev V, Heidenreich A. Diagnosis and management of the growing teratoma syndrome: A single-center experience and review of the literature. Urol Oncol 2018;36:529.
10. Maroto P, Tabernero JM, Villavicencio H, Mesía R, Marcuello E, Solé-Balcells FJ, et al. Growing teratoma syndrome: experience of a single institution. Eur Urol 1997;32:305–9.
11. Denaro L, Pluchinotta F, Faggin R, Manara R, Calderone M, Viscardi E, et al. What's growing on? The growing teratoma syndrome. Acta Neurochir (Wien) 2010;152:1943–6.
12. Huľová S, Aziri R, Chovanec M, Mardiak J, Mego M, Pinďák D. Clinical management and outcome in extreme retroperitoneal growing teratoma syndrome of testicular origin - clinical management and effect of the treatment. Klin Onkol 2019;32:129–32.