CASE REPORT

Primary angiosarcoma of neck nodes with bony metastasis — A case report

Shabbir Hussain Ashiq Ali, Mohammad Jawaid Akhtar Mallick, Ayesha Javed, Abdul Hafeez Lakhan

Abstract

This is a case of a 30 year old female who presented in February 2012 with a large painless left neck swelling since 5 months. Her biopsy was done but the result was inconclusive. Later on after proper staging workup it was found to be bony metastatic disease. Her repeated biopsy showed spindle cell lesion. She was advised surgery, which was done revealing poorly differentiated angiosarcoma involving 27 out of 33 lymph nodes. Angiosarcoma is an uncommon malignant neoplasm characterized by rapidly proliferating, extensively infiltrating anaplastic cells derived from blood vessels and lining irregular blood-filled spaces. Cutaneous angiosarcoma of the scalp and face is the most common form of angiosarcoma. Angiosarcoma of the cervical lymph node without a primary scalp or oral cavity lesion is a very rare presentation and has been reported only once. Thus, this case reported with bony metastasis can be labeled as the first of its kind.

Keywords: Angiosarcoma, Neck nodes, Bony metastasis, Vascular neoplasm.

Introduction

Angiosarcoma is an uncommon malignant neoplasm characterized by rapidly proliferating, extensively infiltrating anaplastic cells derived from blood vessels and lining irregular blood-filled spaces.[1] Specialists apply the term angiosarcoma to a wide range of malignant endothelial vascular neoplasms that affect a variety of sites. They are usually seen in adults, most common sites being the skin, soft tissue, breast, bone, liver and spleen.[2] Angiosarcomas are aggressive and tend to recur locally, spread widely, and have a high rate of lymph node and systemic metastases. The rate of tumour related deaths is high. Approximately 50% of angiosarcomas occur in the head and neck, but they account for less than 0.1% of head and neck malignancies. Cutaneous angiosarcoma of the scalp and face is the most common form of angiosarcoma. The differential diagnosis of angiosarcoma includes haemangioma for the better differentiated lesions, Kaposi's sarcoma for those with a predominantly spindle component, and carcinoma or amelanotic melanoma for the poorly differentiated types.[2]

Department of Radiotherapy, Ziauddin University Hospital, Karachi.
Correspondence: Shabbir Hussain Ashiq Ali.
Email: dr-shabbirhussain@hotmail.com

Case Report

A 30 year old unmarried female reported with a diagnosis of spindle cell neoplasm from her neck node in February 2012. After thorough history and examination it was found that she had complaints of large painless left neck swelling since 5 months. Her initial workup before her presentation here included an FNAC of the neck swelling in posterior triangle, which revealed spindle cell neoplasm with a query of myxofibroma. A trucut biopsy was performed one month later, which again revealed spindle cell neoplasm. After Immunohistochemical staining of the same sample it was then concluded to be a metastatic neuroblastoma. The same sample was then examined in some other institute after a month, which confirmed the earlier findings.

The patient was advised bone marrow biopsy, which proved negative for malignancy. A fresh biopsy was done on the left neck node, which gave the result of 'Poorly Differentiated Malignant Neoplasm'. Bone scan showed metastatic involvement of the right spine of scapula and the T12 vertebra. A CAT scan of neck was then carried out which showed a large soft tissue density mass measuring 12.5 × 11.5 x 8.5 cm in the left side of the neck extending from the subparotid region to the supraclavicular fossa abutting the left lung apex and encasing the bifurcation of external carotid arteries. The mass was compressing the left tonsillar fossa and the base of the tongue. It was also compressing the left lobe of thyroid gland and displacing the trachea towards the right side.

Her ultrasound abdomen showed an echogenic mass measuring 2.1 x 1.1 cm in right lobe of the liver, which was reported as a benign lesion. An extended panel of Immunohistochemical stains were requested on the same sample, which then concluded to be 'Poorly differentiated Synovial Sarcoma'. Surgery was then planned after review by the ENT surgeon. Her Left radical neck dissection was attempted, but was closed by the surgeon without excision of the mass due to profuse bleeding. Surgery was repeated successfully and the histopathology report confirmed it to be a vascular neoplastic lesion, the features of which were consistent with Poorly Differentiated Angiosarcoma with 27 out of 33 lymph

nodes showing involvement by the disease.

Discussion

Angiosarcoma is an uncommon malignant neoplasm characterized by rapidly proliferating, extensively infiltrating anaplastic cells derived from blood vessels and lining irregular blood-filled spaces.[1] The term angiosarcoma applies to a wide range of malignant endothelial vascular neoplasms that affect a variety of sites. These tumours characteristically involve the scalp or face of elderly individuals, where they present as bluish or violaceous plaques and nodules. Most of them spread locally in surface and depth while a third of them eventually give rise to distant metastases, particularly to cervical lymph nodes and lung.[3] Angiosarcomas arising at different sites and in different organs have some distinct features. They may occur in any region of the body but are more frequent in skin and soft tissue and can also originate in the liver, spleen, breast, bone, or heart.

Immunohistochemistry is a powerful tool in the diagnosis of angiosarcoma, particularly in the poorly differentiated histological type. Well known endothelial antigens are F-VIII-RA, CD31, CD34, and Ulex lectin.[4]

Infiltration of bone marrow by angiosarcoma has been reported in literature. K. F. Wong from Hong Kong reported a case of Sinonasal angiosarcoma with marrow involvement, mimicking malignant lymphoma.[5] Chen Wang published another report from University of Toronto with a rare finding of bone marrow involvement in angiosarcoma of the spleen.[6]

All angiosarcomas tend to be aggressive and are often multicentric.[7] These tumours have a high local recurrence rate and metastasis because of their intrinsic biologic properties and because they are often misdiagnosed, leading to a poor prognosis and a high mortality rate.[8] Malignant vascular tumours are clinically aggressive, difficult to treat, and have a reported 5-year survival rate around 20%.[9] Advanced stage at presentation and lack of extensive excision are associated with higher recurrence, higher distant metastasis and low survival rates. The 5-year survival for soft tissue sarcomas is 67%.[10]

The use of irradiation in conjunction with surgery continues to evolve and results in 80% of local control and excellent functional and cosmetic outcome. Considering that 50% of angiosarcomas have distant metastasis, irradiation does not improve survival.

In postoperative therapy, the initial volume is usually treated from 45 to 50 Gy, with subsequent cone downs to a final dose of 60 to 66 Gy, using 1.8 or 2.0 Gy daily fractions. For preoperative irradiation, 45 to 50 Gy is often delivered 2 to 4 weeks before resection with an intraoperative or postoperative boost as indicated by the surgical margin.

Rufus J. Mark collected data of 28 patients of angiosarcomas of the head and neck from 1955 to 1990 in the University of California, Los Angeles (UCLA) and concluded that combined modality therapy offers the best chance for long-term control in patients with angiosarcoma of the head and neck.[11]

Conclusion

This case report gives new information on the occurrence of Angiosarcoma in the cervical lymph node without a primary scalp or oral cavity lesion, which is a very rare presentation and has been reported only once,[10] making this case the first of its kind being reported with bony metastasis.

Disclosures: None.

Conflict of Interest: None.

Funding Source: None.

References

1. Bhattacharya P, Singh P, Bahal A, Gulati D, Tevatia MS, Mehta A, et al. Angiosarcoma of pericardium: a report of two cases. Indian J Pathol Microbiol 2007; 50: 777-9.
2. Juan Rosai. Soft tissues. In: Rosai J. Rosai Ackerman's Surgical Pathology. 9th ed. Missouri: Elsevier; 2009, Pp 2293-4
3. Rosai J, Summer HW, Kostianovsky M, Perez-Mesa C. Angiosarcoma of the skin. A clinicopathologic and fine structural study. Hum Pathol 1976; 7: 83-109.
4. Terada T. Fatal poorly differentiated angiosarcoma of the scalp. Int J Clin Exp Pathol 2010; 3: 541-4.
5. Wong KF, So CC, Wong N, Siu LL, Kwong YL, Chan JK. Sinonasal angiosarcoma with marrow involvement at presentation mimicking malignant lymphoma: cytogenetic analysis using multiple techniques. Cancer Genet Cytogenet 2001; 129: 64-8.
6. Wang C, Rabah R, Blackstein M, Riddell RH. Bone marrow metastasis of angiosarcoma. Pathol Res Pract 2004; 200: 551-5.
7. Keshav P, Hegde SS. Bilateral Primary Angiosarcoma of the Breast. Case Rep Surg 2013; 2013: 139276.
8. Arora D, Mehta Y, Trehan N. Use of recombinant activated factor VII for bleeding following resection of mediastinal angiosarcoma. HSR Proc Intensive Care Cardiovasc Anesth 2011; 3: 197-201.
9. Lezamadel Valle P, Gerald WL, Tsai J. Malignant vascular tumors in young patients. Cancer 1998; 83: 1634-9
10. Weitz J, Antonescu CR, Brennan MF. Localized extremity soft tissue sarcoma: improved knowledge with unchanged survival over time. J Clin Oncol 2003; 21: 2719-25
11. Mark RJ, Tran LM, Sercarz J, Fu YS, Calcaterra TC, Juillard GF. Angiosarcoma of the Head and Neck; The UCLA Experience 1955 Through 1990. Arch Otolaryngol Head Neck Surg 1993; 119: 973-8
12. Yilmaz AS, Oysu C, Tetikkurt S, Semiz-Oysu A. Primary low-grade angiosarcoma of the cervical lymph node. J Otolaryngol Head Neck Surg 2012; 41: E13-5.