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Case Report

Primary Intra-Thoracic Evans Tumor in A Young Female: Case Report and Literature Review

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ABSTRACT

Evans tumor, also known as low-grade fibromyxoid sarcoma, usually presents as a slowly growing, painless mass in young adults, most commonly involving the proximal extremities. Involvement of the intrathoracic compartment has been rarely described, most often as a site of metastasis. Primary intrathoracic low-grade fibromyxoid sarcoma of the pleura is even rarer. Here, we describe a case report of a young lady with a history of psoriatic arthritis who initially presented with shortness of breath due to left pleural effusion. Further workup revealed a primary intrathoracic low-grade fibromyxoid sarcoma of the left pleura.

Keywords: Evans tumor; Fibromyxoid sarcoma; Intrathoracic compartment; Metastasis; Left pleura

Introduction

Low-grade fibromyxoid sarcoma (LGFMS) was first described in 1987 by American pathologist Harry L. Evans and hence, it is also known as Evans tumor. As the name suggests, this tumor is an admixture of cellular myxoid zones and collagenous zones interspersed with bland spindle cells in a whorling or storiform pattern. It is relatively rare and even rarer is when it presents as a primary intrathoracic tumor. Here, we report a case of a primary intrathoracic Evans tumor in a young female.

Case Report

A 37-year-old female with a medical history of psoriatic arthritis (on Ixekizumab) presented to the emergency room with gradual onset dyspnea, easy fatigability and central chest pain, progressively worsening over the last two months. She was a former smoker (2 pack years, quit at age 22) with no major exposures. Admission chest radiograph revealed left-sided pleural effusion, for which thoracentesis was performed, obtaining 800cc of straw-colored fluid. It was transudative by Light's criteria and cytology was consistent with benign mesothelial cells. Further workup included a CT scan of the chest, abdomen and pelvis, which revealed multiple enhancing pleural-based masses involving the left hemithorax (**Figure 1**).

There was no evidence of malignancy in the abdomen or pelvis. Incidentally, serum tumor marker CA125 was elevated. A PET scan revealed mild to moderate FDG-avid left-sided pleural-based masses with calcifications predominantly involving the inferior left hemithorax.

Since the percutaneous ultrasound-guided biopsy was inconclusive and the bronchoscopy was unremarkable, an exploratory left thoracoscopy with biopsy was performed, which revealed multiple pleural-based nodules. The histological specimen consisted of bland spindle cells in a loose myxomatous stroma. Immunostaining was negative for pancytokeratin,

CAM5.2, calretinin and CD34, but positive for MUC4 (**Figure 1**). A diagnosis of primary intrathoracic low-grade fibromyxoid sarcoma was made and immunochemotherapy (Trabectedin/Ipilimumab/Nivolumab) was initiated. Follow up in 6 months documented the stability of the tumour, with no recurrence.

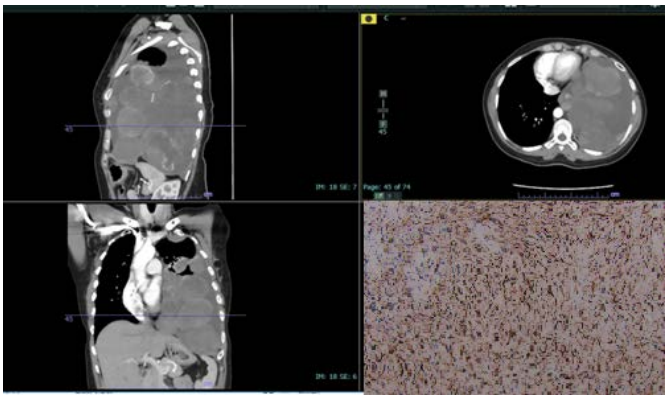

Figure 1: CT Chest in sagittal (upper left), axial (upper right) and coronal views (bottom left) showing a large loculated left pleural effusion containing multiple enhancing pleural-based masses. This is an ill-defined, partially calcified 8.9 x 5.8 cm pleural-based mass (red arrow) and a homogeneously enhancing mass more superiorly (blue arrow) in the pleural space measuring 8.8 x 5.8 cm were noted. Numerous additional morphologically similar masses are present involving the left mediastinal, costal and diaphragmatic pleural surfaces (green arrow). The bottom right image shows strong MUC 4 positive staining (black arrow) on the biopsy specimen, confirming low-grade fibromyxoid sarcoma.

Discussion

Low-grade fibromyxoid sarcoma (also known as Evans tumor) usually appears as a slowly growing, painless mass predominantly in young adults, with a median age of 34 years and a range from 3 to 78 years[1,2]. The most reported site is the proximal extremities, particularly the thigh or trunk originating in the deeper soft tissue layers. Rarely, these tumors have also been found in the viscera, retroperitoneum and intrathoracic compartments, either as a metastatic process or as a primary tumor[3]. These tumors can grow up to 23 cm. The clinical presentation of this tumor varies and can range from gradually progressing respiratory symptoms to a large pleural effusion.

Radiologically, the tumor appears as a multinodular mass with alternating strong (hypercellular) and weak (myxoid) enhancing areas featuring abrupt transitions[5,6]. Ultrasound and MRI findings have also been described[7,8]. Diagnosis is usually made through histology and immunostaining of the tumor after resection.

Histologically, this tumor appears as hyalinizing spindle cells with giant rosettes and fibromyxoid zones. The spindle cells have distinct cytoplasm with a near absence of mitotic figures, hence referred to as "low grade." The fibrous areas have abundant stromal collagen, either as rosettes or with whorled patterns of growth[9,10].

Immunostaining is typically negative for S100, desmin, keratin, pan-cytokeratin, CAM 5.2, calretinin and CD34 and positive for MUC4. LGFMS characteristically expresses the FUS-CREB3L2 fusion (t7;16:q32-34;p11) by RT-PCR[9,11].

Treatment includes excision of the tumor, metastasectomy, radiation and chemoimmunotherapy (trabectedin, ipilimumab, nivolumab)[2,9,11-14].

This tumor may exhibit a progressive course with high rates (6% to 64%) of recurrence (up to 15 years) and metastasis (up to 45 years), showing a "low grade" appearance on histology[4,15]. Although histological appearance should not be used for prognostication, a "de-differentiated recurrence" with anaplastic round cell morphology may indicate short survival. Therefore, it may be necessary to closely follow up with imaging surveillance for a long term after excision[16].

Conclusion

This is a rare case of primary intra-thoracic low-grade fibromyxoid sarcoma in a young female, presenting as pleural based masses. Percutaneous biopsy may be inconclusive and thoracoscopic biopsy may eventually be required. Immunostaining will help in confirming the diagnosis. The treatment involves a combination of surgery, radiation and chemoimmunotherapy.

References

1. Harry L Evans MD. Low-Grade Fibromyxoid Sarcoma: A Report of Two Metastasizing Neoplasms Having a Deceptively Benign Appearance, American J Clin Pathology 1987;88(5):615-619.
2. Cowan ML, Thompson LD, Leon ME, et al. Low-Grade Fibromyxoid Sarcoma of the Head and Neck: A Clinicopathologic Series and Review of the Literature. Head and Neck Pathol 2016;10:161-166.
3. Porrino J, Al-Dasuqi K, Irshaid L, et al. Update of pediatric soft tissue tumors with review of conventional MRI appearance-part 1: tumor-like lesions, adipocytic tumors, fibroblastic and myofibroblastic tumors and perivascular tumors. Skeletal Radiol 2022;51:477-504.
4. Evans Harry L. MD. Low-Grade Fibromyxoid Sarcoma: A Clinicopathologic Study of 33 Cases with Long-Term Follow-Up. The American J Surg Pathology 2011;35(10):1450-1462.
5. Sargar K, Kao SC, Spunt SL, Hawkins DS, Parham DM, Coffin C, Mc Carville MB. MRI and CT of Low-Grade Fibromyxoid Sarcoma in Children: A Report from Children's Oncology Group Study ARST0332. AJR Am J Roentgenol 2015;205(2):414-420.
6. Maeda E, Ohta S, Watadani T, et al. Imaging findings of thoracic low-grade fibromyxoid sarcoma: report of three cases. Jpn J Radiol 2009;27:375.
7. Koh SH, Choe HS, Lee I, et al. Low-grade fibromyxoid sarcoma: ultrasound and magnetic resonance findings in two cases. Skeletal Radiol 2005;34:550-554.
8. Kim SY, Kim MY, Hwang YJ, et al. Low-grade fibromyxoid sarcoma: CT, sonography and MR findings in 3 cases. J Thorac Imaging 2005;20(4):294-297.
9. Mustafa S, VandenBussche CJ, Ali SZ, Siddiqui MT, Wakely PE Jr. Cytomorphologic findings of low-grade fibromyxoid sarcoma. J Am Soc Cytopathol 2020;9(3):191-201.
10. Murshed KA, Al-Bozom I, Ammar A. Sclerosing epithelioid fibrosarcoma: in-depth review of a genetically heterogeneous tumor. APMIS 2021;129(8):455-460.
11. Martínez-Trufero J, Cruz Jurado J, Gómez-Mateo MC, et al. Uncommon and peculiar soft tissue sarcomas: Multidisciplinary review and practical recommendations for diagnosis and treatment. Spanish group for Sarcoma research (GEIS - GROUP). Part I. Cancer Treat Rev 2021;99:102259.

12. Sajid MI, Arshad S, Abdul-Ghafar J, Fatimi SH, Din NU. Low-grade fibromyxoid sarcoma incidentally discovered as an asymptomatic mediastinal mass: a case report and review of the literature. J Med Case Rep 2021;15(1):50.

13. Demetri GD, von Mehren M, Jones RL, et al. Efficacy and Safety of Trabectedin or Dacarbazine for Metastatic Liposarcoma or Leiomyosarcoma After Failure of Conventional Chemotherapy: Results of a Phase III Randomized Multicenter Clinical Trial. J Clin Oncol 2016;34(8):786-793.

14. D'Angelo SP, Mahoney MR, Van Tine BA, et al. Nivolumab with or without ipilimumab treatment for metastatic sarcoma (Alliance A091401): two open-label, non-comparatives, randomized, phase 2 trials. Lancet Oncol 2018;19(3):416-426.

15. Saab-Chalhoub MW, Al-Rohil RN. Low-grade fibromyxoid sarcoma of acral sites: Case report and literature review. J Cutan Pathol 2019;46:271-276.

16. Folpe AL, Lane KL, Paull G, Weiss SW. Low-grade fibromyxoid sarcoma and hyalinizing spindle cell tumor with giant rosettes: A Clinicopathologic study of 73 cases supporting their identity and assessing the impact of high-grade areas. Am J Surg Pathol 2000;24(10):1353-1360.