

# An autopsy study of synchronous intratesticular and mediastinal sarcoma with pulmonary metastasis

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## ABSTRACT

Most testicular sarcomas arise from germ cell tumors (GCTs) viz. teratoma or spermatocytic seminoma. Pure intratesticular sarcomas are very rare. We present an autopsy report of non-germ cell primary intratesticular sarcoma with synchronous mediastinal sarcoma and lung metastasis in a young male who presented with symptoms of mediastinal compression and simultaneously had a significant gradually enlarging testicular mass.

**Key words:** Intratesticular, lung metastasis, mediastinum, sarcoma

## INTRODUCTION

Intrascrotal sarcomas are divided into testicular and paratesticular sarcomas. Attempts have to be made to delineate the specific site of origin of the sarcoma. Since these structures are in continuity with each other it is not always possible to determine the exact origin in such infiltrative tumors.<sup>[1,2]</sup> Paratesticular sarcomas refer to tumors which involve epididymis, scrotal wall, and spermatic cord. Intratesticular sarcomas are far less common, and comprise less than 1%. Majority of them, in reality are an element of germ cell neoplasia.<sup>[1-3]</sup> Pure intratesticular non-germ cell sarcomas are extremely rare with very few cases reported in the literature.<sup>[1-8]</sup> To our knowledge, synchronous pure intratesticular and mediastinal sarcoma has not been reported as yet.

## CASE REPORT

A 38-year-old male was admitted for progressive breathlessness, cough with mucoid expectoration,

and intermittent chest pain for past 6 months, with a history of waxing and waning fever for 15 days. His physical examination revealed an enlarged left testis. The patient did not disclose about the testicular mass because it was painless, gradually progressing over a year. Chest X-ray and computed tomography (CT) chest showed right sided pleural effusion with a pleura-based mediastinal mass measuring 12 cm × 8 cm × 4 cm. Cytological study of pleural fluid was positive for malignant cells. Lung was biopsied, but the biopsy was non-representative. He succumbed on the 6<sup>th</sup> day of hospitalization. Autopsy examination revealed 20 cc of turbid hemorrhagic fluid in the thoracic cavity. There was a large, irregular, gray tan left-sided mediastinal mass measuring 12 cm × 10 cm × 8 cm [Figure 1]. Cut sections of the mass showed fleshy appearance. The adhered lower lobe of left lung showed a distinct nodule measuring 2 cm × 2 cm. Other lung appeared unremarkable. Left testicle was enlarged with a bosselated appearance measuring 12 cm × 12 cm × 7 cm. Cut section showed a tumor mass with nodules showing areas of hemorrhage and extensive necrosis [Figure 2]. Contralateral testis was normal. There was no iliac lymphadenopathy. Histopathological examination of testicular mass showed a tumor composed of spindle cells arranged in a fascicular pattern [Figure 3a], hemangiopericytomatous pattern [Figure 3b], myxoid areas with cells arranged in reticular pattern [Figure 3c], and focal chondroid

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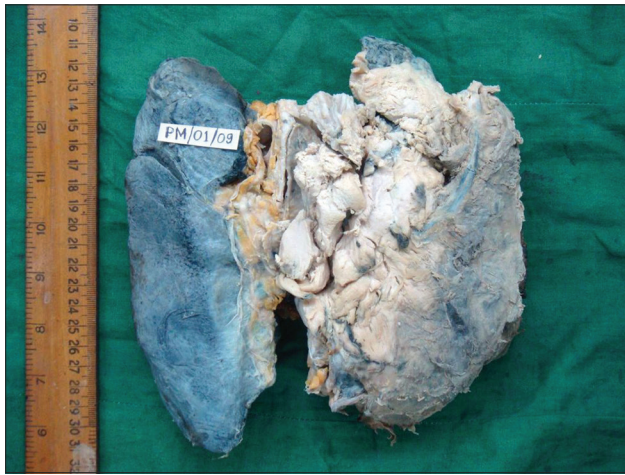
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area [Figure 3d]. Interspersed pleomorphic bizarre tumor giant cells [Figure 4], extensive necrotic areas, and brisk mitoses were evident. Thorough sampling and extensive search did not show any teratomatous or seminomatous element to exclude the germ cell origin of the tumor. Histopathological sections from the mediastinal mass and lung nodule showed exactly similar morphology like testicular tumor with fascicular arrangement of spindle cells, myxoid areas, and reticular pattern along with bizarre giant cells but devoid of cartilaginous foci. A panel of immunohistochemistry (IHC) including vimentin, cytokeratin, desmin, myogenin, smooth muscle actin (SMA), placental alkaline phosphatase (PLAP), S100, c-Kit, and CD30 was run. Vimentin was positive [Figure 5]. All other markers were negative. We rendered a diagnosis of pure intratesticular sarcoma and synchronous mediastinal sarcoma with lung metastases.



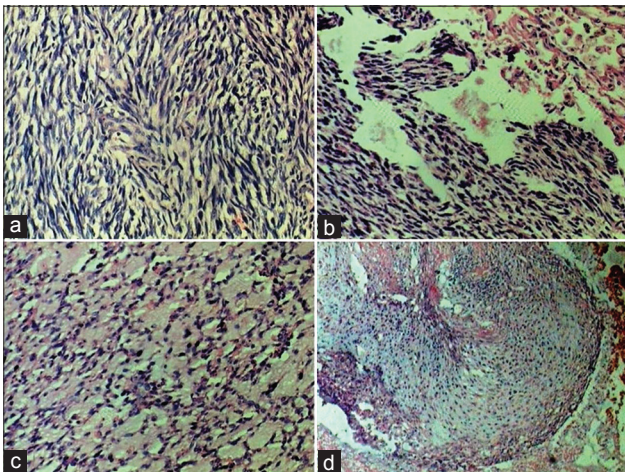
**Figure 1:** Gross photograph showing lungs with large, irregular, grey tan mediastinal fleshy mass encasing the left lung

## DISCUSSION

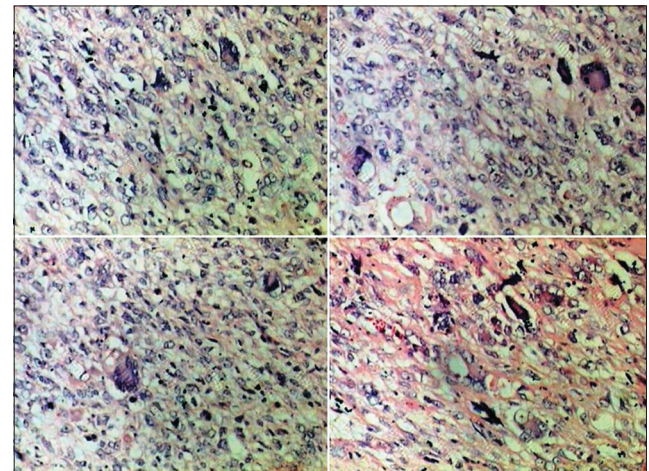
Primary intratesticular sarcoma is a unique tumor with little information in the literature about its natural history and treatment protocol due to its rarity.<sup>[1]</sup> Mediastinal masses present earlier due to compressive symptoms. Sarcoma of the testis usually presents as a painless swelling.<sup>[5]</sup> Our index case, presented with symptoms of compression but had a simultaneous involvement of testis and mediastinum. He did not give relative importance to the testicular mass as it was slow growing and painless. Testicular mass was evident only on physical examination. Morphologically, we found unclassified sarcoma of testis. In such cases, primary testicular sarcoma, germ cell tumor (GCT) with sarcomatous component (SC), and paratesticular rhabdomyosarcoma (RMS) are the tumors under differential diagnosis.<sup>[6,7]</sup> Paratesticular tissue was free, hence, paratesticular sarcoma was ruled out. However, no GCT component was seen in multiple sections studied, hence, GCT with SC was ruled out. GCTs of ovary,



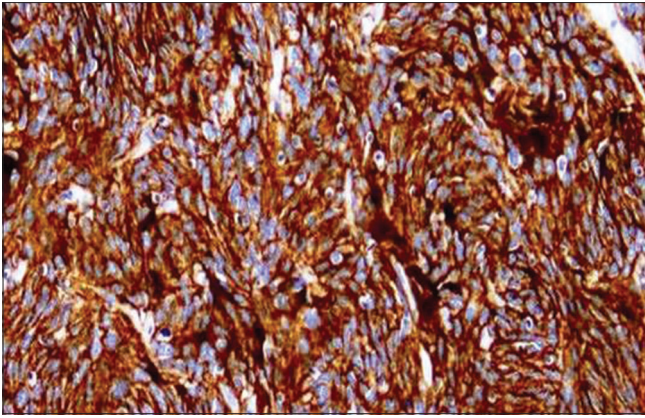
**Figure 2:** Testis replaced by a fleshy nodular tumor mass with extensive areas of necrosis (H and E,  $\times 10$ )



**Figure 3:** (a-d) Histopathological sections showing testicular tumor composed of spindle cells arranged in a fascicular pattern (H and E,  $\times 10$ ), hemangiopericytoma-like pattern (H and E,  $\times 10$ ), myxoid areas with cells arranged in reticular pattern (H and E,  $\times 10$ ), and focal chondroid area (H and E,  $\times 40$ )



**Figure 4:** High power view showing pleomorphic bizarre tumor giant cells (H and E,  $\times 40$ )



**Figure 5:** Immunohistochemical study showed diffuse vimentin positivity

mediastinum, testis, and intracranial cavity may develop as a SC,<sup>[4]</sup> although it is relatively common in mediastinal GCTs, it is rare, accounting for 3-6% in testicular GCTs.<sup>[1]</sup> IHC helped us confirm our diagnosis of pure intratesticular sarcoma by a negative desmin, myogenin, and SMA which ruled out smooth muscle origin, negative c-Kit ruled out seminoma, negative PLAP ruled out teratoma and negative CD30 ruled out embryonal carcinoma. The metastasis of sarcomatous component of testicular GCT to mediastinum is a well-known phenomenon.<sup>[4,5]</sup> However, we could not establish whether testicular sarcoma was primary or secondary involvement as the deceased had neglected the testicular enlargement. Lung metastases appeared to be due to be contiguous spread from mediastinal tumor. RMS, spindle cell sarcoma, fibrosarcoma, osteosarcoma, leiomyosarcoma, chondrosarcoma, Kaposi sarcoma, and unclassified sarcoma of the testis have been reported.<sup>[1-8]</sup> Very few pure testicular sarcomas have been reported in the literature. Sarcomas are derived from the supporting cells of testicular interstitium: Peritubular myoid cells, endothelial cells, smooth muscle cell, or primitive mesenchymal cell. There is lack of data on the natural history, histological criteria for diagnosis and treatment recommendations because of rarity of this disease. Most cases reported in the literature indicate that intratesticular sarcoma may be an indolent tumor with a potential for cure if treated early.<sup>[5]</sup> Adjuvant chemotherapy with single-agent adriamycin or adriamycin plus ifosfamide has been the mainstay of treatment for advanced and metastatic soft tissue sarcomas from other sites.<sup>[8,9]</sup> The same regimens can be used as palliative therapy for testicular and mediastinal sarcomas. On this premise, our patient was put on adriamycin, but

he succumbed on the very next day. Rarity of the condition and hence, lack of literature makes it difficult to define and optimize therapy.<sup>[1-9]</sup>

## CONCLUSION

To conclude, pure intratesticular sarcomas are very rare and a thorough search for germ cell element is essential before diagnosing a pure intratesticular sarcoma. Concurrent mediastinal sarcoma may appear with earlier manifestations due to compressive symptoms.

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