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Paratesticular Osteosarcoma—A Rare Tumor with Distinctive Imaging Findings

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Abstract

Extraosseous osteosarcoma (EOO) is a rare mesenchymal malignancy representing 4% of all osteosarcomas and 1% of soft tissue sarcomas. The testes, its supporting structures, that is, paratestes, and the spermatic cord are among the rarest sites for EOO, with only 11 published English language reports to date. We report our experience with a 73-year-old male presenting with left hemiscrotal swelling, noted to have extensive amorphous intratumoral calcification on imaging. He underwent left high inguinal orchidectomy with en bloc hemiscrotectomy, with a final pathologic diagnosis of primary paratesticular osteosarcoma. Our literature review corroborates this distinctive, hitherto overlooked imaging feature.

Keywords
- osteosarcoma
- paratestis
- calcification
- spermatic cord
- orchidectomy

Introduction

Extraosseous osteosarcoma (EOO), also known as soft tissue osteosarcoma, refers to osteosarcoma occurring in soft tissue rather than bone. EOOs are uncommon, accounting for only 4% of all osteosarcomas and 1% of soft tissue sarcomas.[1] Osteosarcoma arising from the testis, its supporting structures, that is, paratestis or spermatic cord are exceedingly rare, with only 11 published English language reports to date. In this report, we present a 73-year-old male presenting with left hemiscrotal swelling, noted to have extensive amorphous calcification on imaging. Extirpative pathology revealed primary paratesticular osteosarcoma. We also review published literature with particular emphasis on imaging characteristics.

Case History

A 73-year-old patient presented with progressively enlarging left hemiscrotal mass of 2 months duration. He denied history of trauma. Physical examination revealed a 15 cm bony hard mass in the left hemiscrotum infiltrating overlying skin. The left testis was palpable separately at the inferior pole of the mass. Contralateral testis was normal. Physical examination was otherwise unremarkable. Serum tumor markers (alpha fetoprotein, beta-human chorionic gonadotropin and lactate dehydrogenase) were normal. Ultrasonogram of the scrotum revealed a 10 cm mass with heterogenous echotexture and multiple calcifications. Computed tomography (CT) revealed an ill-defined heterogeneously enhancing mass with extensive amorphous intratumoral calcifications (►**Fig. 1**). Both testes

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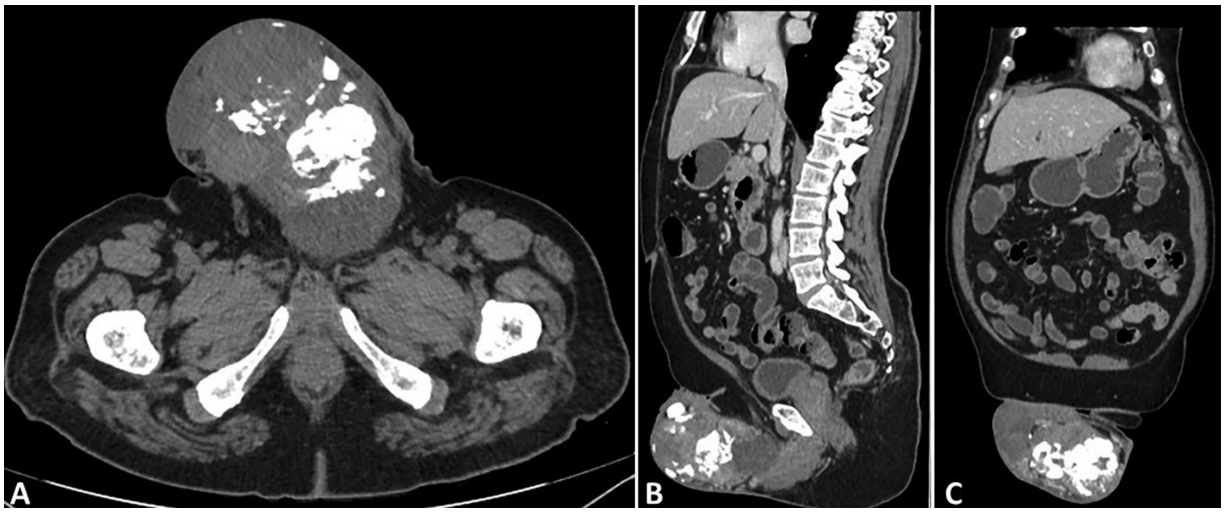

Fig. 1 Computed tomographic images showing a large left paratesticular tumor with extensive amorphous calcification. (**A**) Cross-sectional imaging, (**B**) sagittal image, and (**C**) coronal image through the tumor.

were noted separate from the mass and appeared normal. Metastatic evaluation was negative. With a provisional diagnosis of malignancy, we performed a metastatic evaluation, including a CT of the chest, abdomen, and pelvis, which was negative.

He underwent left high inguinal orchidectomy with en bloc hemiscrotectomy. Intraoperatively, a discrete bony hard swelling, with the left testis adherent to its lower pole, was noted. Gross pathology showed a $16.5 \times 10 \times 7.8$ cm variegated tumor adjacent to the testis, with cut surface showing solid and cystic areas. Calcified areas and necrosis were also seen. Histology revealed spindle to polyhedral cells with scant eosinophilic cytoplasm and hyperchromatic nuclei, with 14 mitoses per 10 high power field. Foci of osteoid formation, calcification, and osteoclast-like multinucleated giant cells were also noted (►**Fig. 2**), suggesting a diagnosis of high-grade osteosarcoma arising from the paratestis. This was corroborated on immunohistochemistry, which was positive for vimentin, variable positivity for CD 99, and negative for SATB2, S100, desmin, CD34, and keratin.

After discussion in a multidisciplinary tumor board, considering the lack of evidence for benefit of adjuvant therapy in localized disease, the patient was advised surveillance. He remains disease free at 8 months of follow-up.

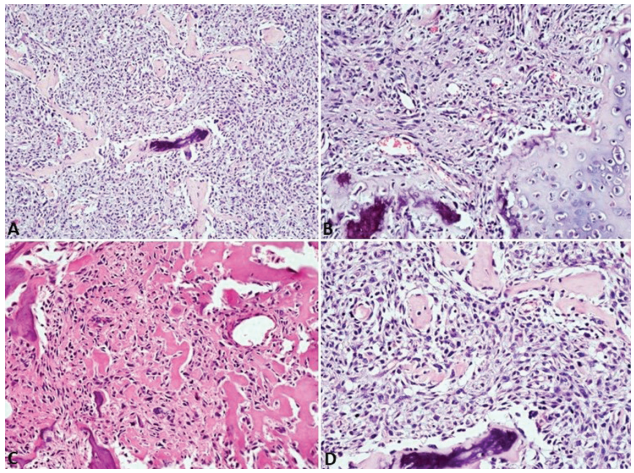

Fig. 2 Histopathology. (**A**) Sheets of tumor cells with intervening osteoid matrix undergoing focal mineralization (hematoxylin and eosin [H&E 100X]). (**B**) H&E 200X. (**C**) Tumor cells with hyperchromatic pleomorphic nuclei with adjacent osteoid and chondroid matrix (H&E 200X). (**D**) Spindle-shaped tumor cells with hyperchromatic, pleomorphic nuclei with intervening osteoid matrix following decalcification (H&E 200X).

Discussion

EOOs are rare malignant mesenchymal neoplasms that produce osteoid, bone, and occasionally cartilage. To be characterized as EOO, these tumors must (i) have a unified sarcoma pattern (excluding mixed malignant mesenchymal tumor), (ii) produce bone-like and/or cartilage matrix, (iii) be primarily located in soft tissues without skeletal attachment, and (iv) a metastasis from a primary skeletal osteosarcoma elsewhere must be excluded.[1] The soft tissues of the lower followed by the upper extremity are most commonly affected, and visceral involvement is rare. To the best of our knowledge, only 11 published reports in English language, of EOO of the testis, paratestis, or spermatic cord exist (►**Table 1**).

EOO is hypothesized to arise from neoplastic transformation of sequestered primitive mesenchymal cells or embryonic osteogenic tissue. In the testis, EOO must be distinguished from sarcomatous transformation of germ cell tumors by excluding the presence of the later in the resected specimen by meticulous pathologic examination, as was performed in our case.

Based on the available data, common clinical presentation in the fourth to eight decades of life, with a large, hard, hemiscrotal or groin mass, that developed over the course of a few weeks to months (►**Table 1**).

No pathognomic imaging features have been described to date. We reviewed all published reports, with particular attention to findings on imaging studies. At presentation, the tumor tends to be large with longest tumor diameter ranging between

Table 1 Summary of published reports of testicular, paratesticular, and spermatic cord osteosarcoma

	Study	Year	Age at presentation (years)	Primary organ of origin	Time to presentation (weeks)	Tumor LTD (cm)	Calcification on Imaging	Treatment	Adjuvant therapy	Outcome	Follow-up duration (months)
1	Mathew et al[2]	1981	73	Testis	8	3.5	NA	Orchidectomy	None	No recurrence	2.5
2	Zukerberg et al[3]	1990	30	Testis	52	6.0	NA	High inguinal orchidectomy	None	No recurrence	66
3	Lee et al[4]	2004	78	Testis	2	4.0	Calcified mass	High inguinal orchidectomy with RPLND	None	No recurrence	44
4	Tazi et al[5]	2006	60	Testis	16	12.0	Calcified mass	High inguinal orchidectomy	None	No recurrence	16
5	Resorlu et al[6]	2018	63	Testis	NA	16.0	NA	High inguinal orchidectomy	None	No recurrence	12
6	Al-Masri et al[10]	2007	52	Paratestis	16	17.0	NA	High inguinal orchidectomy	Adjuvant cisplatin, doxorubicin, and methotrexate	Expired due to retroperitoneal metastasis	6
7	Hong et al[4]	2012	52	Paratestis	52	6.5	Calcified mass	High inguinal orchidectomy	None	No recurrence	9
8	Spirtos et al[7]	1991	55	Spermatic cord	1	4.0	NA	High inguinal orchidectomy	None	No recurrence	24
9	Beiswanger et al[5]	1997	54	Spermatic cord	NA	7.0	Calcified mass	Wide local excision + high inguinal orchidectomy	None	No recurrence	9
10	Stella et al[6]	2007	59	Spermatic cord	260	25.0	Calcified mass	High inguinal orchidectomy	Inguinal and pelvic radiation (60Gy)	Expired due to lung metastasis at 9 years	132
11	Ugidos et al[11]	2010	62	Spermatic cord (liposarcoma + osteosarcoma)	NA	9.0	NA	High inguinal orchidectomy	None	No recurrence	8

Abbreviations: LTD, longest tumor dimension; NA, not available; RPLND, retroperitoneal lymph node dissection.

3.5 and 25.0 cm (►**Table 1**). Modest heterogenous enhancement is usually observed. Imaging is often remarkable for extensive amorphous intratumoral calcification. This likely results from ossification by osteosarcoma tumor cells. Although observed in previously published reports, these unique imaging finding have not been not emphasised.[4–8] Calcification may occur in other testicular tumors such as nonseminomatous germ cell tumors, particularly teratomas, and nongerm cell tumors such as large-cell calcifying Sertoli cell tumor. However, this is usually focal and/or heterogenous and not as pronounced as seen in osteosarcoma.[10,12] Calcification may also be noted in long-standing hydrocele or hematocele, in which case it is usually peripheral and shell like.[13]

Unlike EOO at other sites which commonly present with metastatic disease,[1] majority of reported EOO arising from testis, paratestis or spermatic cord have presented with localized disease. As with our patient, surgical resection, comprising high inguinal orchidectomy with wide local excision of the tumor has been the mainstay of management in most reports. Due to rarity of disease, the value of adjuvant chemotherapy, radiation, or prophylactic retroperitoneal lymphadenectomy in localized disease remains unknown. However, despite the lack of any adjuvant treatment, regional or metastatic recurrence has been reported in only two instances,[9,14] suggesting a favorable prognosis in majority of patients with localized disease.

Conclusion

Although EOOs of testis, paratestis, or spermatic cord are exceedingly rare, they should be included in the differential diagnosis of a male in the fourth to eight decades of life presenting with a hard hemiscrotal or groin mass with extensive amorphous intra-tumoral calcification on imaging. Surgical resection remains the mainstay of management.

Note

The study was conducted at the Cancer Institute Chennai, India.

Funding

None.

Conflict of Interest

None declared.

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