Rhabdomyosarcomatous differentiation in a spermatocytic seminoma with review of literature

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ABSTRACT

The sarcomatous differentiation occurring in spermatocytic seminoma (SS) renders an aggressive behavior with metastatic potential to this relatively indolent neoplasm. Correct identification of this sarcomatous component is essential as further management differs. Herein, we report a case of young male with SS with rapid increase in size of the tumor. Histopathology revealed a rhabdomyosarcomatous component infiltrating the rete-testis and epididymis along with a well-circumscribed SS.

Key words: Rhabdomyosarcomatous differentiation, spermatocytic seminoma, testis

INTRODUCTION

Spermatocytic seminoma (SS) is an uncommon testicular germ cell neoplasm with distinct histopathological features. It usually occurs in older men with mean age of 53.6 years (range 19-72 years). It bears an excellent prognosis but tends to behave aggressively and renders a poor prognosis if associated with sarcomatous transformation. Patients with sarcomatous transformation at the time of presentation already have metastatic disease and this is almost fatal. The sarcoma component may present as rhabdomyosarcoma, angiosarcoma, leiomyosarcoma, myxoid liposarcoma or undifferentiated sarcoma. The identification and diagnosis of the sarcomatous component is pertinent for planning further management. We describe a case of a young man with SS developing rhabdomyosarcomatous transformation.

Clinical findings

A 38-year-old man presented with long standing history of swelling in right hemiscrotum. He underwent right testicular biopsy in a local medical college, which was diagnosed as seminoma. The patient was referred to our institute for further evaluation. On examination, he gave a history of sudden increase in size of the mass. There was a right testicular mass measuring 10 × 10 cm, with scrotal violation and left testis was normal. His general physical examination was normal. The serum tumor markers i.e., alpha-fetoprotein (AFP) and beta HCG were within normal. Hematological and biochemical parameters were normal. Chest X-ray, CECT abdomen and pelvis were normal. He underwent right high inguinal orchidectomy and hemiscrotectomy in view of scrotal violation. Retroperitoneal lymph node dissection (RPLND) was not performed as radiologically there was no lymphadenopathy. In addition to local and abdominal radiotherapy, chemotherapy (with vincristine, actinomycin D and cyclophosphamide) was given. Two months postoperatively, the patient is on follow-up and doing well.

Pathological Findings

The right high inguinal orchidectomy specimen measured 7 × 6 × 4 cm with attached vas deferens and scrotal skin. The testicular shape was maintained. On cut surface, a tumor measuring 6 × 6 cm was seen replacing the entire testicular parenchyma. The tumor was yellow, soft, and homogenous with a lobulated appearance. Most of the nodules were gelatinous with some appearing darker in color and firm in consistency than the rest [Figure 1a]. Microscopically, the spermatocytic component consisted of polymorphic cell population with large cells having...
characteristic ‘spermine’ chromatin; intermediate-sized cells and small lymphocyte-like cells [Figure 1b]. The mitotic activity was very brisk [Figure 1c]. At places, the cells were clustered around the vessels. The cells were immunopositive with c-kit (CD117) [Figure 1d] but were negative with PLAP (placental-like alkaline phosphatase). The other component of the tumor consisted of fascicles of spindle shaped cells with fusiform nuclei [Figures 2a and b]. The sarcomatous component was seen destroying the epididymis and rete testis. Admixed with both the components were many multinucleated and bizarre tumor giant cells with hyperchromatic nuclei. Few strap cells and globoid cells were identified within the fascicles of spindle cells [Figure 2c]. No morphologic transition between the two cell types was apparent. No areas of intratubular germ cell neoplasia (ITGCN) or other associated germ cell components were identified. The tunica albuginea was, however, not infiltrated by the tumor. No lymphovascular emboli were seen. Immunohistochemically, the spermatocytic component was negative for placental alkaline phosphatase (PLAP), CD30, smooth muscle-actin, and the spindle cells were strongly positive for desmin [Figure 2d] and weakly for myoglobin.

DISCUSSION

SS is a relatively rare, well-defined pathological entity first described by Masson\[5\] comprising 3-7% of all seminomas.\[3\] It is an indolent neoplasm with long duration of symptoms, early stage at presentation, absence of metastasis and bears an excellent prognosis.\[6\] However, dedifferentiation or transformation to a sarcoma which occurs in approximately 6% of cases renders a poor prognosis with a metastatic potential.\[7\] Literature review cites small series\[3\] and a few case reports\[8-10\] of occurrence of sarcomatous differentiation in SS. The occurrence of rhabdomyosarcomatous differentiation is rare and has been described in the literature in eight previous case reports [Table 1].\[6,8,9,11\] In the present case, the sarcomatous transformation occurred at a much early age of 38 years compared to the previous cases. The sudden increase in size should always suggest emergence of a sarcomatous component. Light microscopic findings are usually diagnostic after one appreciates the two components distinctly. However, it is important to be aware of misinterpreting the focal areas of atypical stromal overgrowth within conventional germ cell tumor. True sarcomatous component can be distinguished by its expansile growth pattern, infiltration into the surrounding elements and characteristic morphology of the distinct sarcomatous components.\[13\] Immunohistochemistry helps in highlighting and further categorizing the sarcomatous element.

The seminomatous component was well circumscribed but the rhabdomyosarcomatous component was seen infiltrating and destroying the rete testis and epididymis. Identification of epithelial structures of rete testis is important as these may be mistaken for epithelial component of a mixed nonseminomatous germ cell tumor.

The origin of the sarcomatous elements of these tumors and the relation to the spermatocytic component is uncertain. Few have implied\[12,13\] this sarcoma component to be a teratomatous element of a mixed germ cell tumor as the occasional occurrence of sarcomatous elements (chiefly rhabdomyosarcoma, angiosarcoma, and leiomyosarcoma) in gonadal and extragonadal germ cell tumors is well known. The serum elevation of alpha-fetoprotein and
beta-HCG favors this hypothesis. The serum tumor markers were not elevated in the present case. However, other authors view the sarcomatous component as an ‘anaplastic transformation’ or ‘dedifferentiation’ occurring within the well-differentiated neoplasm which seems most plausible.\textsuperscript{[3]} The development of this tumor, first with slow enlargement of the testis followed by rapid increase in size tends to favor the explanation of sarcomatous differentiation occurring in SS.

The treatment of choice in SS is orchidectomy, followed by surveillance to detect tumor in the contralateral testis. However, in the presence of sarcomatous differentiation, adjuvant chemotherapy or radiotherapy is beneficial. As the risk of metastases if remarkably increased with this transformation, a close follow-up is essential; however, prognosis is dismal despite aggressive multimodality approach.\textsuperscript{[3,6]} In conclusion, this case describes the rare sarcomatous differentiation occurring in SS heralded by a rapid increase in size, which portends a poor prognosis to this indolent neoplasm. Accurate and timely diagnosis is pertinent for planning further management of the patient.

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### Table 1: Review of reported cases of spermatocytic seminoma with rhabdomyosarcomatous differentiation

| Authors          | Age  | Gradual enlargement | Rapid phase | Size              | Involvement at orchidectomy | Histology of sarcoma | Follow-up                                      |
|------------------|------|---------------------|-------------|-------------------|----------------------------|----------------------|------------------------------------------------|
| Matoska et al.\textsuperscript{[4]} | 55 years | 2 years            | 1 month     | 9×6×3 cms         | Testis                    | Rhabdomyosarcoma     | Died in 14 months, pulmonary and liver metastasis |
| Matoska et al.\textsuperscript{[4]} | 56 years | 5 years            | 5 months    | 9×6×3 cms         | Testis                    | Rhabdomyosarcoma     | Died in 15 months, pulmonary metastasis          |
| Matoska et al.\textsuperscript{[4]} | 60 years | -                  | -           | 25×20×12 cm       | Extratesticular and distant metastasis | Rhabdomyosarcoma     | Died in one month, lymphnode, lung, thyroid, heart metastasis |
| Matoska et al.\textsuperscript{[4]} | 51 years | 2 years            | -           | 18×12×12 cm       | Testis, extratesticular and distant metastasis | Rhabdomyosarcoma     | Died in 2 months, lungs, liver retroperitonea metastasis |
| True et al.\textsuperscript{[3]}   | 60 years | -                  | 3 months    | 25×20×15 cm       | Testis, extratesticular and distant metastasis | Rhabdomyosarcoma     | Diagnosed at autopsy                             |
| Menon et al.\textsuperscript{[9]}  | 55 years | 10 years           | 7 months    | 15×9×8 cms        | Testis                    | Rhabdomyosarcoma     | Patient was non compliant, follow up not available |
| Robinson et al.\textsuperscript{[11]} | 44 years | 18 months         | 1 month     | 17 cms            | Testis                    | Rhabdomyosarcoma     | Died in 5 months                                 |
| Chelly et al.\textsuperscript{[6]}  | 50 years | 4 months           | -           | 14 cms            | Testis                    | Rhabdomyosarcoma     | Not mentioned                                   |
| Present case     | 38 years | 2 years            | 2 months    | 7×6×4 cms         | Testis                    | Rhabdomyosarcoma     | Doing well 2 months postoperative               |
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