Primary testicular lymphoma with rupture: An unusual presentation

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Abstract

Primary testicular lymphoma usually presents as a unilateral testicular mass with occasional bilateral involvement. The tumor show contiguous spread to rete testis, epididymis spermatic cord and rarely to tunica albuginea. We report a case of primary testicular

lymphoma which showed rupture of tunica albuginea with involvement of inguinal lymph node which is unusual. A 50-year-old male patient presented with right inguinal swelling and right side scrotal swelling of five months' duration. Fine needle aspiration of the right inguinal lymph node was done and was suggestive for lymphoma/seminoma. Histopathology of right orchiectomy revealed non-Hodgkin's lymphoma (NHL). Further investigations did not reveal any other organs involved with non-Hodgkin's lymphoma. Primary testicular lymphoma usually shows spread to extranodal sites like skin, central nervous system and Waldeyer's ring at presentation and at relapse. Whereas, less common sites are lung, bone, liver, gastrointestinal system and nodal sites, especially the paraaortic lymph nodes. Testicular lymphoma with involvement of the inguinal lymph node is unusual. Clinical presentation of such cases may mimic germ cell tumors.

Key words: *Lymphoma, primary, testis*

INTRODUCTION

Lymphoma of the testis comprises 3-5% of all testicular tumors.[1] Testicular lymphoma is an aggressive extranodal lymphoma that arises primarily in the testis or as a part of generalized NHL. Primary testicular lymphoma (PTL) usually presents as a unilateral testicular mass of variable size and can show contiguous spread to rete testis, epididymis, spermatic cord and rarely to tunica albuginea.[2] They can frequently involve extranodal sites like skin, CNS and Waldeyer's ring at presentation and at relapse.[2] Less common sites are the lung, bone, liver, GIT and nodal sites, especially the paraaortic lymph nodes.[2] Testicular tumor metastasizing to the inguinal node is unusual. We present a case of PTL which showed involvement of the tunica albuginea with rupture and rare involvement of inguinal lymph node.

CASE REPORT

A 50-year-old male presented with a right side testicular swelling and right side inguinal lymph node enlargement since five months. Fine needle aspiration cytology of the inguinal lymph node showed cells having anisokaryosis, round vesicular nucleus, irregular nuclear border/nuclear clefts with scant cytoplasm. A diagnosis of NHL/metastatic deposits from testis was suggested.

We received the orchiectomy specimen measuring 8 × 6 × 5 cm. External surface showed a raw area/breach in capsule measuring 6 × 5 cm. Cut section showed a well-circumscribed grey-white tumor measuring 6 × 3 cm with focal areas of hemorrhage [Figure 1]. Periphery of the tumor showed normal compressed testicular tissue. Spermatic cord measured 9 × 2 cm. Cut section was unremarkable.

Microscopy showed a small round monotonous population of cells showing isokaryosis and scanty cytoplasm, arranged in sheets [Figures 2 and 3]. The stroma in between showed scanty fibrovascular tissue. Amidst the tumor tissue were seen a few eosinophils. Areas of confluent necrosis were also seen. Differential diagnoses of seminoma and NHL were offered. Immunohistochemistry showed that the tumor cells were positive for CD 20 [Figure 4], and negative for CD 5, CD23 and Cyclin D1. Mib 1 proliferation index was 90%. There was immunoexpression for both kappa and lamda light chain. A final diagnosis of high-grade NHL of B-cell phenotype was considered.

The patient was then subjected to chest X-ray, ultrasound, CT scan, bone marrow and cerebrospinal fluid examination to look for involvement of the CNS, other lymph nodes and organs by NHL. There was no evidence of NHL elsewhere in the abdomen. The HIV status of the patient was negative. Hence a final diagnosis of primary testicular lymphoma with spread to the inguinal lymph node was considered.

DISCUSSION

PTL is an aggressive extranodal lymphoma and is usually seen in the elderly age group, but recently it has shown an increased incidence in the younger age group.[3] This could be attributed to the increased prevalence of HIV infection.[3] Another potential cause for incidence in younger age, could be that the immunophenotypic characterization of testicular tumors in recent years has led to a better recognition of PTL and less frequent misinterpretation as germ cell tumor.

PTL usually presents as a unilateral testicular mass of variable size. Grossly, lymphoma is seen as a solid, homogenous, grey-white mass with a lobulated appearance replacing the testis. Bilateral involvement can also occur at presentation and has been reported in 18% of cases.[4] The tumor usually also shows contiguous spread to the tunica albuginea, rete testis, epididymis and spermatic cord. In one large series, involvement of the epididymis and spermatic cord was seen in 60% and 39% of cases respectively whereas involvement of the tunica albugenia is very rare.[2]

Histologically, diffuse large B-cell lymphoma (DLBCL) is by far the most common type of NHL. Other reported types include follicular lymphoma, plasmacytoma, and lymphoblastic and burkitt-like lymphomas.[2] DLBCL shows obliteration of testicular parenchyma by neoplastic

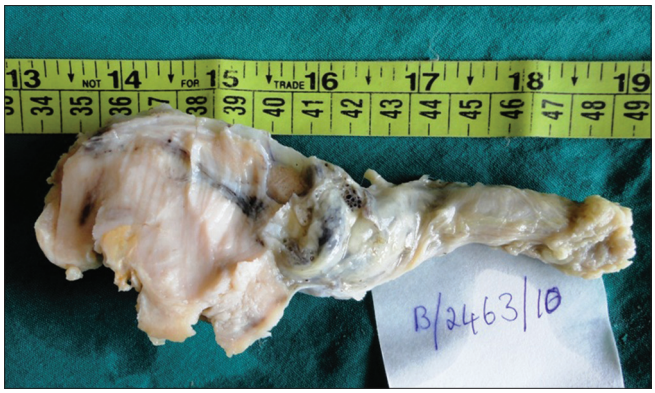

Figure 1: Grey-white tumor with adjacent compressed testis and epididymis. Tumor shows a breach of the tunica albuginea at one end

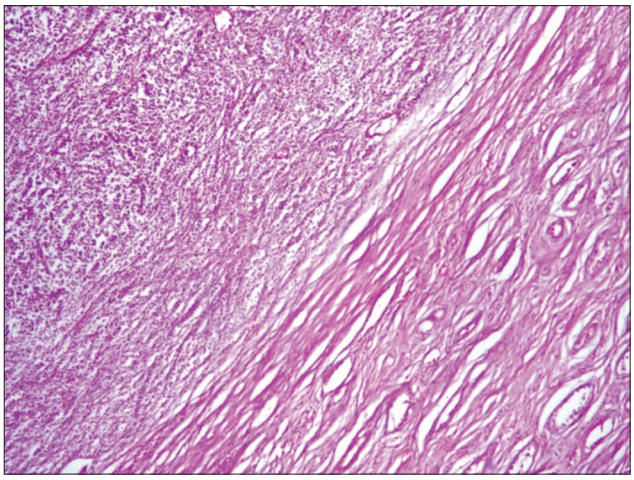

Figure 2: Monotonous population of tumor cells with adjacent testicular tissue (H and E ×40)

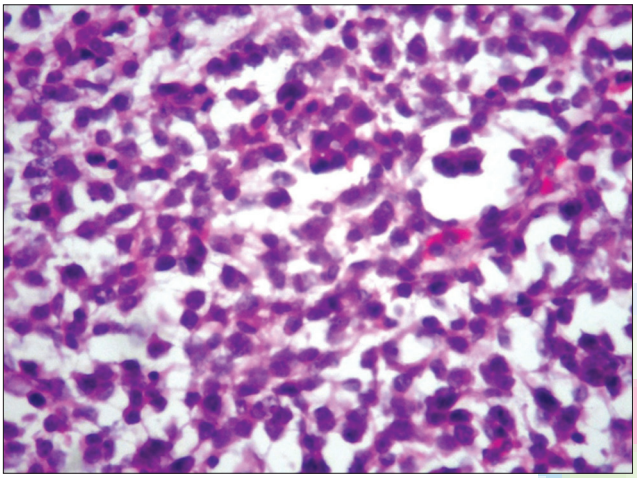

Figure 3: Tumor cells showing isokaryosis and scant cytoplasm separated by thin fibrovascular septa (H and E ×400)

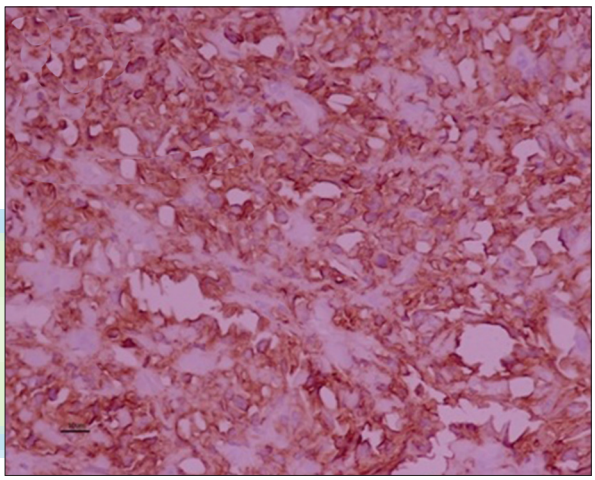

Figure 4: Immunohistochemistry showing tumor cells positive for CD20

cells arranged in solid sheets and separated by thin fibrous tissue. At the periphery, the tumor may show a distinctive intertubular growth pattern with splaying of seminiferous tubules by irregular aggregates, clusters and cords of tumor cells. The tumor cells are large with ill-defined cell membrane, and variable amounts of non-vacuolated cytoplasm. Nuclei are pleomorphic with irregular and twisted nuclear borders, fine chromatin and sometimes inconspicuous nucleoli. Lymphocytes, plasma cells, eosinophils and histiocytes may be seen, especially at the periphery of the tumor. Single-cell necrosis may be seen, but large foci of necrosis are much less frequent.[5]

The major mimickers of PTL are seminoma and embryonal carcinoma. Seminomas show tumor cells arranged in sheets with fibrous septa in between showing lymphocytic infiltration. The tumor cells are usually large, have a distinct cell membrane with abundant clear cytoplasm with a centrally placed large nucleus and one to two prominent nucleoli. Embryonal carcinoma typically has a more pleomorphic appearance with more abundant cytoplasm and prominent areas of hemorrhage and necrosis.[5] Immunohistochemistry is essential in differentiating these tumors, as treatment options in these entities also vary.

Recently, immunohistochemistry has led to further classification of DLBCL into two groups—non-germinal center type which carries a poor prognosis than the germinal center type which carries a better prognosis.[6] The two different categories can be identified by the expression pattern of CD10, Bcl-6 and MUM1. Germinal-center DLBCL expresses CD10 and Bcl-6. Non-germinal center DLBCL is negative for CD10 and positive for MUM1. The majority (89%) of primary DLBCL belong to the non-germinal center type and have a high proliferative activity.[6]

Testicular lymphoma has an aggressive clinical course and can frequently involve extranodal sites at presentation and at relapse.[2] PTL is reportedly associated with involvement of the skin and subcutaneous tissue in 6-13%, Waldeyer's

ring in 4-6% and CNS in 3-6%. The CNS and Waldeyer's ring are common sites of involvement at relapse and carry a poor prognosis. Less common sites are the lung, bone, liver, GIT and nodal sites, especially the paraaortic lymph nodes.[2]

Testicular tumor metastasizing to the inguinal node is unusual, and is possible only when there is scrotal involvement. The primary path for testicular cancer metastasis is to the retroperitoneal lymph nodes. The inguinal lymph nodes are not usually involved, unless the tumor has invaded the scrotum, or a scrotal incision was made during biopsy or orchiectomy, or there was prior scrotal surgery such as repair of a hydrocele or a varicocele. In the present case, we believe that inguinal node involvement could be because of a rupture of testicular lymphoma which has spread to the parietal layer of the tunica vaginalis and scrotum, which is an unusual case. The patient was subjected to chemotherapy and follow-up for three months, wherein the inguinal swelling reduced in size. However, the patient was lost for further follow-up.

Rupture of lymphoma has been reported commonly in the spleen and is sometimes the presenting symptom.[7] Rare sites of rupture include the small intestine and heart.[8] An extensive search of the literature did not reveal any case of testicular lymphoma with rupture. However, one case of testicular lymphoma with inguinal node swelling without other site involvement has been reported, but the reason for metastasis has not been discussed.[9] The present case emphasizes an unusual presentation of testicular rupture with inguinal node involvement.

REFERENCES

1. Seymour JF, Solomon B, Wolf MM, Janusczewicz EH, Wirth A, Prince HM. Primary large cell non hodgkin's lymphoma of the testis: A retrospective analysis of patterns of failure and prognostic factors. Clin Lymphoma 2001;2:109-15.
2. Ferry JA, Harris NL, Young RH, Coen J, Zietman A, Scully RE. Malignant lymphoma of testis, epididymis and spermatic cord: A clinicopathologic study of 69 cases with immunophenotypic analysis. Am J Surg Pathol 1994;18:376-90.
3. Wilson WT, Frenkel E, Vuitch F, Salgowsky AI. Testicular tumors in men with human immunodeficiency virus. J Urol 1992;47:1110-1.
4. Hurley LJ, Burke CR, Shetty SK, Previte SR, Sakr OE, Libertino JA. Bilateral primary non Hodgkin's lymphoma of the testis. Urology 1996;47:596-8.
5. Abaddi MA, Hattab EM, Tarawneh M, Orazi A, Ulbright TM. Primary testicular and paratesticular lymphoma. A Retrospective Clincopathologic Study of 34 cases with emphasis on differential diagnosis. Arch Pathol Lab Med 2007;131:1040-6.
6. Abaddi MA, Hattab EM, Tarawneh M, Amr SS, Orazi A, Ulbright TM. Primary testicular diffuse large B-cell lymphoma belongs to the nongerminal center B–cell like subgroup. A study of 18 cases. Mod pathology 2006;19:1521-7.
7. Chappuis J, Simoens C, Smets D, Duttman R, Mendes De Costa P. Spontaneous rupture of the spleen in relation to a Non Hodgkin's lymphoma. Acta Chir Belg 2007;107:446-8.
8. Kataoka I, Arima F, Nishimoto J, Watanabe T, Kobayashi Y, Tamura R, *et al*. Enteropathy – type T cell lymphoma showing repeated small bowel rupture and refractoriness to chemotherapy: A case report. Jpn J Clin Oncol 2002;32:546-9.
9. Balogun TM, Omodele FO, Olaiya MA. Primary lymphoma of testis in remission for more than ten years: A case report. West Afr J Med 2009;28:387-90.

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