Case Report

Primary extranodal marginal zone lymphoma – Epididymis

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ABSTRACT

An elderly male presented with painful swelling in the right side of scrotum. He was treated with antibiotics for epididymoorchitis without any response. Ultrasound examination revealed a hypoechoic vascular mass in the tail of the epididymis. Fine needle aspirate cytology was inconclusive. Excision of the mass was done and biopsy revealed primary extranodal marginal zone lymphoma arising from mucosa associated lymphoid tissue (MALT) of epididymis. Marginal zone lymphoma arising from the MALT of epididymis is very rare. Lymphoma should be considered as a differential diagnosis of any epididymal swelling unresponsive to conservative treatment. We report a rare case of primary extranodal marginal lymphoma of MALT arising from epididymis.

Key words: B-cells, epididymis, lymphoma, marginal zone

INTRODUCTION

Primary extranodal marginal zone lymphomas (MZL) of mucosa associated lymphoid tissue (MALT) are commonly seen in the gastrointestinal tract. It also been reported to arise from genitourinary organs like kidney,[1] urinary bladder,[2] and prostate.[3] We report a rare case of primary extranodal MZL of MALT arising from epididymis.

CASE REPORT

A 70 year-old male presented with painful swelling in the right scrotum of 9 months duration. His past medical history was negative for tuberculosis or trauma. Earlier he was treated as chronic epididymoorchitis elsewhere and no regression of symptoms was noted. On examination, a firm well-defined tender swelling of 3 × 2.5 cm was palpable in right epididymal region separately from the testis. All the blood and urine investigations were normal. Ultrasonogram of scrotum revealed a well-defined hypoechoic paratesticular lesion of 2.1 × 1.9 cm size with increased vascularity [Figure 1] in relation to tail of right epididymis. Both testes were normal. Ultrasound examination of the abdomen was normal. FNAC of the right epididymal mass was done and it showed few lymphoid infiltrate with atypical features, but was inconclusive. Right radical orchiectomy was planned. Intraoperatively, a well-encapsulated grayish-white mass of 2.5 × 2 cm was noted to arise from the lateral aspect of right epididymis close to its tail [Figure 2]. Right testis and rest of the cord structures were normal. The mass was excised along with a segment of epididymis. Right testis was preserved. Gross specimen examination showed a nodular mass of 2.5 × 2 cm size. Cut section of the mass revealed a vaguely lobulated grayish-white surface with scattered few microcystic areas. Microscopy revealed that excised segment of epididymis was replaced by a tumor composed of ill-defined nodules and sheets of medium-sized lymphoid cells with round to oval nuclei having finely clumped chromatin, inconspicuous nucleoli, and scanty cytoplasm [Figure 3]. Few interspersed larger cells with centrally placed nucleoli were seen. Few reactive follicles were also seen. The lymphoid cells were positive for CD20 [Figure 4], bcl-2, CD43, and negative for CD5, CD10, and CD23. Mib-1 proliferation index was low (<5%) except in reactive follicles. The features were suggestive of low-grade non-Hodgkin lymphoma of extranodal marginal zone type. Bone marrow aspirate and trephine biopsy were done and did not show evidence of lymphoma involvement. Blood

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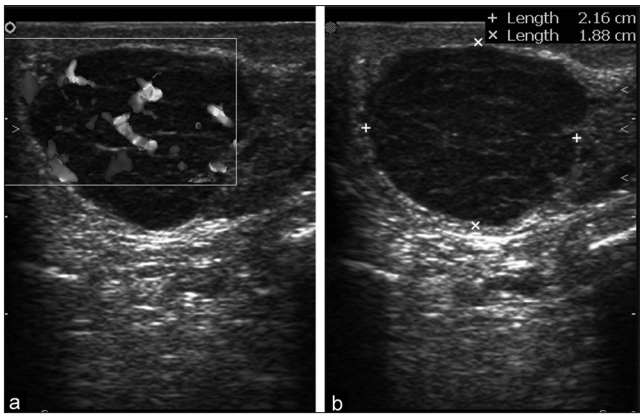

Figure 1: Well-defined hypoechoic paratesticular lesion seen in relation to the tail of right epididymis with increased vascularity

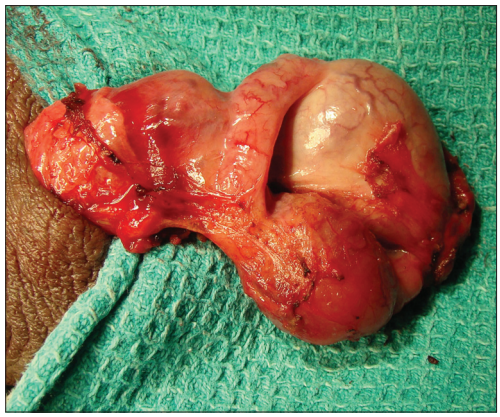

Figure 2: Well-encapsulated lesion seen replacing the segment of epididymis close to the globus minor

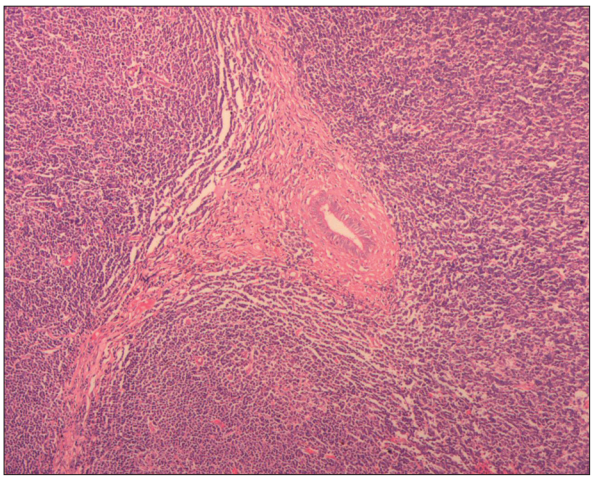

Figure 3: (H and E, ×200) Dense lymphoid infiltrate obscuring the epididymal architecture with occasional remnant epididymal tubules

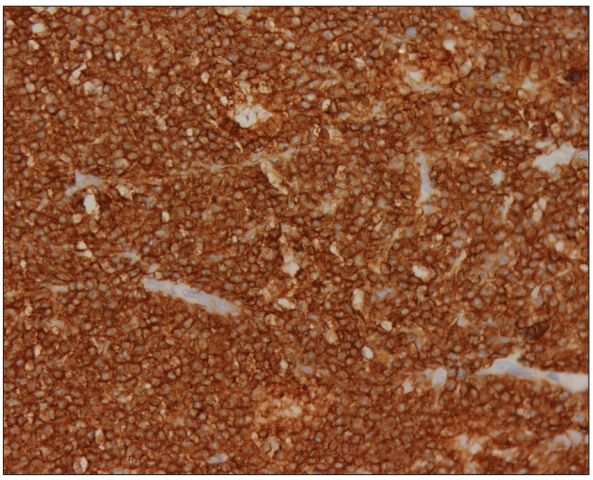

Figure 4: (CD20 immunostain, ×400) The lymphoid cells showed diffuse membrane staining for CD20

borne virus screening was negative. The staging work-up indicated a diagnosis of stage IE (extranodal) primary MALT lymphoma of epididymis. The postoperative period was uneventful. He was advised for surveillance in view of the low-grade lymphoma. At 14 months of follow-up, no recurrence was noted.

DISCUSSION

MZL are believed to arise from B-cells present in the marginal zone, which is the zone outer to the mantle zone of B-cell follicles. The group of MZL comprises of three entities namely: splenic MZL, nodal MZL, and extranodal MZL of MALT.[4] MALT lymphoma usually arises in mucosal sites where lymphocytes are not normally present and where MALT is acquired in response to either chronic infectious conditions or autoimmune processes: *Helicobacter pylori* gastritis, Hashimoto's thyroiditis, Sjogren syndrome.[5]

Extranodal MZL of MALT type has been described in most of the organs. In urinary organs, it has been described in kidney, prostate, and urinary bladder. The histological features of extranodal MZL of MALT type are similar regardless of site of origin. A very occasional case of primary MZLs of MALT type arising from epididymis has been reported.[6] Possible factors predisposing to the MZL formation in epididymis is not known. Lymphomas of the epididymis present with painful or painless localized swelling of the epididymis. They are often diagnosed as chronic or atypical epididymoorchitis, tuberculosis, and paratesticular benign or malignant tumors. Constitutional symptoms are extremely uncommon. The diagnosis is often established by the biopsy of the epididymal mass. The B-cells of MALT lymphoma show the immunophenotype of the normal marginal zone B-cells present in spleen, Peyer's patches, and in lymph nodes. Therefore, the tumor B-cells express surface immunoglobulins and pan-B antigens (CD19, CD20, and CD79a) and lack CD5, CD10, CD23, and cyclin D1 expression.[7] The diversity of subtypes affirms the importance of fully characterizing lymphomas by immunohistochemistry and other modalities, which are indispensable for accurate diagnosis. There is no protocol available for the management of MZL arising from the epididymis as the disease condition is very rare. Radical

orchidectomy was done for the only case reported.[6] As the disease is a low-grade lymphoma, the treatment can be tailored according to each individual case. When an epididymal mass does not benefit from medical treatment, lymphoma should be considered as a differential diagnosis. Up to our knowledge, this is the second case of rare primary extranodal MZL arising from the epididymis reported in literature.

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