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Case Report

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Primary (poorly differentiated) sclerosing liposarcoma of the temporal region. An uncommon tumor in a rare site: A case report

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ABSTRACT

Liposarcoma (LS) in the head and neck region is a rare tumor. The sclerosing variant of LS is a subtype of well-differentiated LS characterized by areas of conventional LS admixed with hypocellular areas of stromal sclerosis that show atypical lipomatous cells. The (poorly differentiated) sclerosing LS, on the other hand, is more cellular with atypical, pleomorphic and often bizarre giant tumor cells admixed with atypical lipoblasts. We report a case of poorly differentiated sclerosing LS of temporal region in a 49-year-old man. Radiologically, the tumor was dumbbell shaped with intra and extra cranial extension. In this case, we discuss the clinico-radiological and pathological findings of an unusual tumor in a rare location.

KEY WORDS: Sclerosing liposarcoma, temporal region, S100

INTRODUCTION

Liposarcoma's (LS) are one of the most common soft tissue tumors, being second only to Pleomorphic sarcoma (earlier called malignant fibrous histiocytoma). However, its occurrence in the head and neck region is rare, accounting for around 5% of the cases. Only two cases of LS occurring in the temporal region have been reported previously. Of the several variants recorded, the poorly differentiated sclerosing type is a recently reported entity [1]. The clinical presentation, morphological features and diagnostic approach are discussed.

CASE REPORT

A 49-year-old woman presented with slowly growing, painless swelling in right temporal region of 4 months duration. Computed tomography (CT) scan showed a well-defined heterogeneously enhancing mass lesion showing focal areas of fat density within, extending from the right infratemporal fossa up to the lateral orbital margin. The mass was extending superiorly along the temporalis muscle and laterally extending superior to and beyond the zygomatic arch up to the skin. Medially the mass was abutting the lateral pterygoid muscle. There was no evidence of any underlying bony erosion or destruction seen [Figure 1]. A clinical impression of soft tissue tumor, possibly sarcoma was made. Per-operatively the lesion was avascular with good plane and capsule between lesion and surrounding tissue. Excised tumor was 55 g and 10×4×4cm in dimensions. On cut section, tumor had yellowish grey with multilobulated appearance and focal avascular myxoid areas.

The histopathological sections showed a tumor composed of broad bands of sclerosed stroma with widely dispersed pleomorphic spindle cells, scattered bizarre giant cells, and adipose tissue with areas of myxoid change and scattered multivacoulated pleomorphic lipoblasts [Figure 2a and b]. Immunohistochemically, tumor cells were desmin negative with scattered S-100 positive cells [Figure 3]. Based on these findings, the tumor was reported as poorly differentiated Sclerosing LS.

A 2 years follow-up of the patient was uneventful.

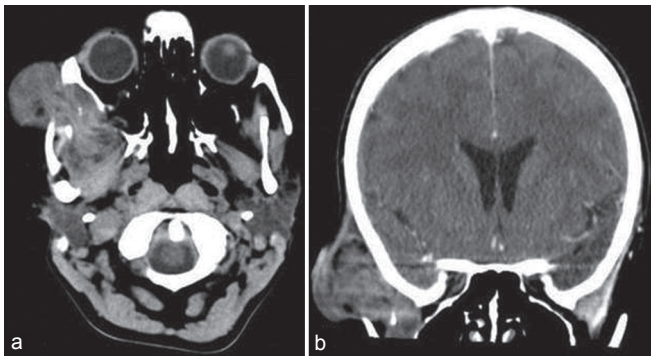

Figure 1: Axial (a) and coronal (b) contrast enhanced computed tomography brain: A well-defined heterogeneously enhancing mass lesion showing focal areas of fat density seen in the right temporal and infratemporal region.

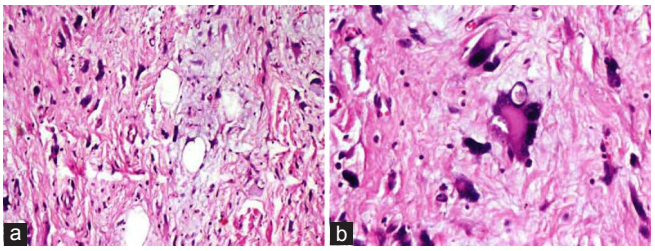

Figure 2: (a) Section shows a tumor composed of sclerotic stroma with scattered pleomorphic lipoblasts (H&E, ×200). (b) Scattered multinucleate, bizzare, tumor giant cells were seen (H&E, ×400).

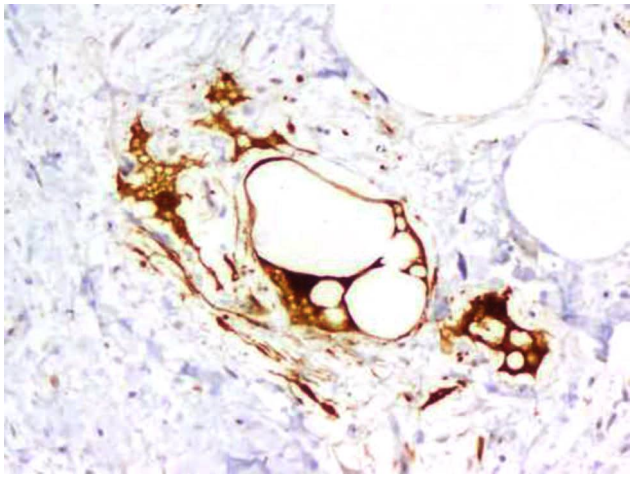

Figure 3: S100 immunohistochemistry showing nuclear and cytoplasmic positivity in spindle shaped and bizarre multivacuolated lipoblasts (S100, ×400).

DISCUSSION

Being the second most common malignant soft tissue tumor, LS accounts for 10-35% of all soft tissue sarcomas [2]. It occurs predominantly in the extremities and retroperitoneum. Accounting for 5% of head and neck sarcomas, [2] common sites include the larynx, hypopharynx, oral cavity, orbita, scalp and soft tissues of the neck [3]. Only 2 cases have been previously reported to occur in the temporal region [4].

The rarity of these tumors and lack of information regarding the clinico-pathological features and prognostic factors further hinders early diagnosis and treatment planning. Clinically they present as soft to firm, painless soft tissue masses varying in size depending upon the site of involvement. They can be easily mistaken clinically as lipoma, cyst or other benign soft tissue tumors. They do not generally arise from pre-existing lipomas [3].

Radiologically, the presence of adipose tissue within the tumor mass is a clue to the diagnosis of LS. Well-differentiated tumors exhibit a diagnostic appearance on CT or magnetic resonance (MR) images, with a largely lipomatous mass (±75% of the lesion) and non-lipomatous components with thick septa or focal nodules. The CT or MR imaging finding of a nodular dominant focus of non-lipomatous tissue in a well-differentiated LS suggests dedifferentiation. The high water content of myxoid LS is reflected on imaging studies. Pleomorphic LS are heterogeneous tumors with small amounts of fat seen on MR images in 62-75% of cases [2]. This was observed in the present case as well.

Majority of the cases of LS are well-differentiated (lipoma-like). Several histological variants have been described, including myxoid, sclerosing, inflammatory, epithelioid, pleomorphic, spindle cell, round cell and dedifferentiated LS [1]. The World Health Organization however recognizes only five distinct histologic subtypes of LS's: Well differentiated, dedifferentiated, myxoid, pleomorphic, and not otherwise specified. The "mixed" subtype described in the previous classification has been removed [5]. The sclerosing variant is generally regarded as a subtype of well-differentiated LS with areas of conventional lipoma-like LS admixed with hypocellular areas of stromal sclerosis that may contain atypical lipomatous cells. This variant is most often seen in the retroperitoneum and paratesticular regions [6]. The present case was unusually located extracranially in the right temporal region.

Suster and Morrison [1] in their series of 8 cases described LS with a proliferation of highly atypical, pleomorphic lipoblastic cells along with extensive areas of sclerosis (30-50%) and called them as poorly differentiated sclerosing type LS; these tumors were associated with a poorer prognosis when compared with well differentiated LS's. The present case also showed similar pleomorphism. The presence of marked atypia excludes a well differentiated Sclerosing LS and the lipoblasts seen in this case rule out dedifferentiated LS. This tumor is purported to arise de-novo or in a pre-existing lipoma. This case may have perhaps originated *de-novo*, in the absence of history of a pre-existing swelling in the same location.

Immunohistochemically the tumor cells in well-differentiated areas may be positive with S-100 protein, however this may be lost in poorly differentiated and dedifferentiated areas. The role of immunohistochemistry remains largely to exclude other malignant mimics which include - pleomorphic sarcoma, parachordoma etc.

In the absence of distinctive immunohistochemical markers coupled with the non-availability of specialized cytogenetic and molecular assays in most practice settings, morphological criteria form the basis for the diagnosis of LS. The demonstration of the classical multivacuolate lipoblasts with eccentric indented, hyperchromatic nucleus and lipid rich cytoplasm is decisive.

Cytogenetic study helps in this regard with the presence of supernumerary 'ring' and/or giant rod chromosomes containing amplified segments from the 12q13-15 region and MDM2 associated co-amplification being decisive [7].

Wide surgical excision is the treatment of choice for LS to obviate the likelihood of satellite nodules and hence increased recurrence. Lymph node dissection is not indicated unless there is concrete evidence of metastasis. Nonsurgical treatment modalities are of limited use in LS [3]. Post-operative radiotherapy is useful for localized lesions, while chemotherapy is recommended for systemic disease. Our patient was managed with complete surgical excision and did not receive post-operative radiotherapy.

To conclude, poorly differentiated sclerosing LS is an unusual variant of LS with atypical manifestations and an aggressive outcome. An uncommon entity in the maxillofacial location it requires preoperative and radiological assessment, wide excision and long-term monitoring.

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