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Case report

US and CT findings of multicentric leiomyosarcomatosis

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Abstract. This article presents a case of leiomyosarcomatosis with widespread lesions involving the soft tissues and the most unlikely organs such as thyroid and salivary glands, pancreas, ligamentum teres, bladder wall, and bones without lymph node or distant metastasis. The CT and US findings of this rare phenomenon are discussed with regard to the literature.

Key words: Leiomyosarcoma – Diagnosis – Ultrasound – Computed tomography

Introduction

Leiomyosarcoma (LMS) is an uncommon malignant tumor of smooth muscle origin, which tends to arise mostly in gastrointestinal tract, retroperitoneum, urinary tract, uterus, and soft tissue [1]. Here we present a case of leiomyosarcomatosis with widespread lesions involving the most unlikely organs such as thyroid and salivary glands, pancreas, ligamentum teres, and bones.

Case report

A 65-year-old female patient presented with a complaint of painless swellings in her body. Incisional biopsy taken from the left deltoid and right intercostal muscles revealed whitish tissues. Histologic, microscopic, and immunohistochemical studies showed spindle cell sarcoma of smooth muscle origin with high mitotic activity, moderate pleomorphism, and prominent muscle fascicles. With the diagnosis of leiomyosarcoma, US and CT of the neck, thorax, and abdomen were performed in order to show the spread of the disease.

Ultrasound scans of the neck showed multiple hypoechogenic, well-defined nodules 5–20 mm in size in the tongue, thyroid gland, and submandibulary glands (Figs. 1–3). Also the same type of lesions 10–15 mm in size were observed within the right sternocleidomastoid muscles, suprahyoid muscles, and left masseteric muscle on both US and CT scans (Fig. 4). The nodule in the masseter, unlike the ones described above, was necrotic and larger in size. The CT scans of the neck revealed an enhancing soft tissue mass destroying the left ramus mandibula in the left masseter (Fig. 4b).

Ultrasound scans of the abdomen showed hypoechogenic nodules at the head and corpus of the pancreas, ligamentum teres, and at the perivesical area indenting the anterior wall of the urinary bladder, 1–2 cm in size (Figs. 5, 6). Nodular involvement of the anterior abdominal wall was easily detected with US, however, and paravertebral muscles were also observed. Thoracal and abdominal contrast-enhanced CT scans showed multiple nodules involving the additional involvement of the intercostal, paravertebral, iliopsoas, and gluteal muscles (Fig. 7). Apart from the lesions defined with US scans, CT showed a soft tissue mass destroying the T4 vertebra and extending into the spinal canal (Fig. 8). Multinodular involvement of the pleura was also observed (Fig. 9). There was also a moth-eaten lytic lesion at the right iliac bone (Fig. 10). All of the lesions, within the limits of color Doppler US scan, also had central increased vascularity with high resistive indices (RI) which were almost always greater than 0.60. No lymphadenopathy was detected.

Considering the localization of the lesions and the absence of involvement of the lymph nodes or the parenchyma of the lung or the liver, these tumors could not be attributed to metastasis. Multicentric synchronous leiomyosarcomatosis seemed to be the most likely diagnosis. Fine-needle aspiration biopsy of the nodules localized in the thyroid and the submandibular glands was also compatible with LMS.

Discussion

Leiomyosarcoma is a relatively uncommon malignant mesenchymal tumor that exhibits smooth muscle differentiation [2]. It is a tumor of adult life and seen more

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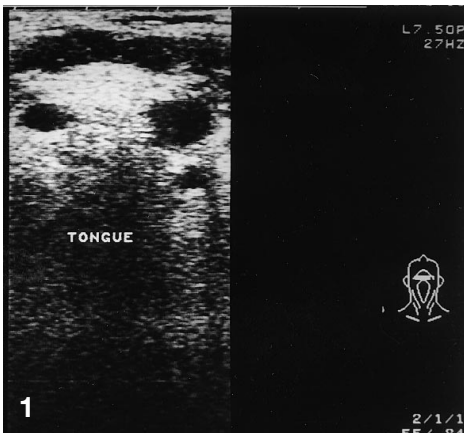
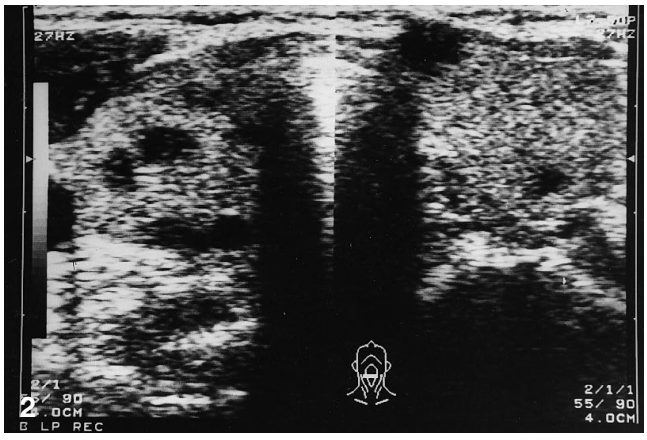
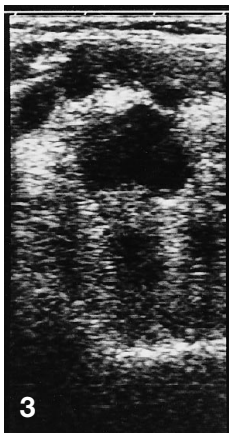
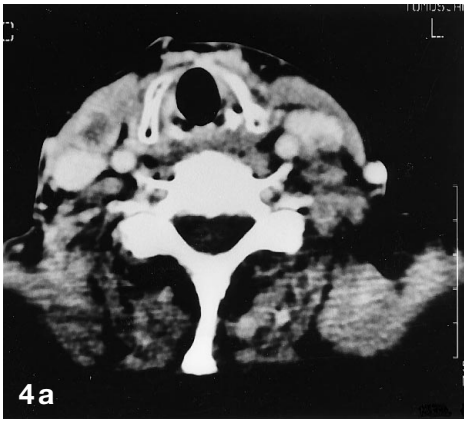








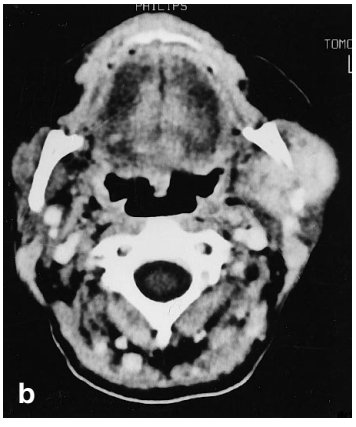


Fig. 1. Ultrasound scan of the mouth floor reveals two clearly defined hypoechogenic nodules in the tongue *(TONGUE)*. The rest of the normal tongue parenchyma is hyperechogenic

Fig. 2. Ultrasound scan of the thyroid reveals bilateral multiple hypoechogenic nodules < 10 mm in size. The size of the thyroid gland was normal

Fig. 3. Ultrasound scan of the left submandibular gland shows multiple hypoechogenic nodules enlarging the gland

Fig. 4 a, b. Axial CT scans of the neck. **a** A hypodense nodular lesion 10–15 mm in size with peripheral contrast enhancement enlarges the right sternocleidomastoid muscle. **b** An enhancing soft tissue mass destroying the left ramus mandibula is seen in the left masseteric muscle

commonly in women than in men, comprising only 7% of all sarcomas [3]. Generally, it has a poor prognosis with high-percentage recurrence and hematogenous metastasis especially to the lung, bone, and brain [2, 4].

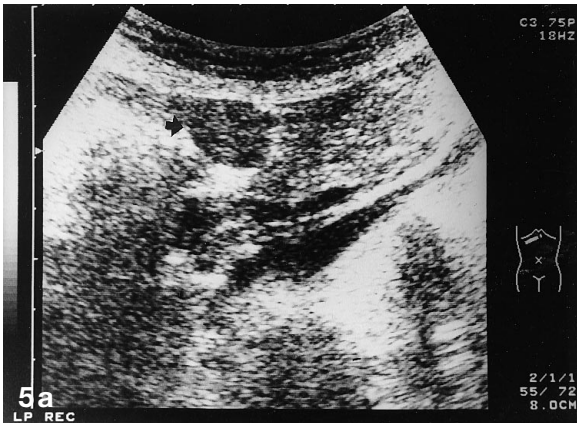
Leiomyosarcoma arises in head and neck in only 3–10% of cases, and of these, paranasal sinuses (19%), cervical esophagus (12%), and skin and soft tissues (16%) are the most common locations [3]. Jaw bones are the predominant localization of oral involvement. Primary LMS of the tongue is very rare, and up to now only 6 cases have been reported in the international literature [3, 5, 6]. In contrast, metastatic involvement of the mandible for bony deposits and the gingiva and tongue for soft tissue deposits are more common [2]. Leiomyosarcoma involving the salivary glands is an extremely uncommon phenomenon [4]. Only two cases of parotid-gland LMS have been reported in the literature thus far, and except for our case, to our knowledge, there is no other report showing submandibular gland infiltration.

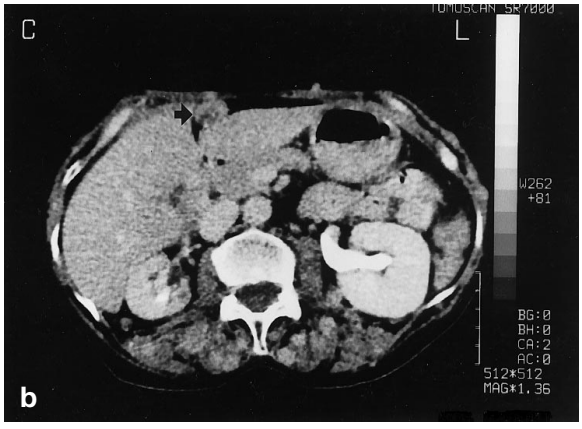
Primary smooth muscle tumors of the thyroid gland are infrequent [7]. Primary LMS of the thyroid tends to be a large mass with necrosis and hemorrhage, invading the extrathyroidal tissue. To our knowledge, only one case of primary LMS of the thyroid presenting as multiple nodules has been reported in the international literature [7].

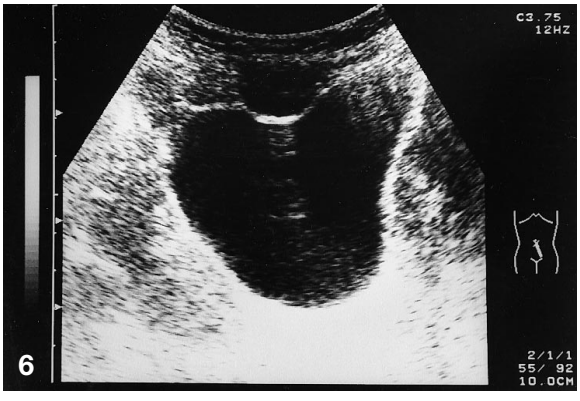
Pancreatic LMS intrinsic to the gland is postulated to be derived from vascular or ductal smooth muscles [8]. Radiologic differentiation of these tumors from other neoplasms of the pancreas is impossible, since they mimic both the primary and secondary neoplasms of the pancreas [9].

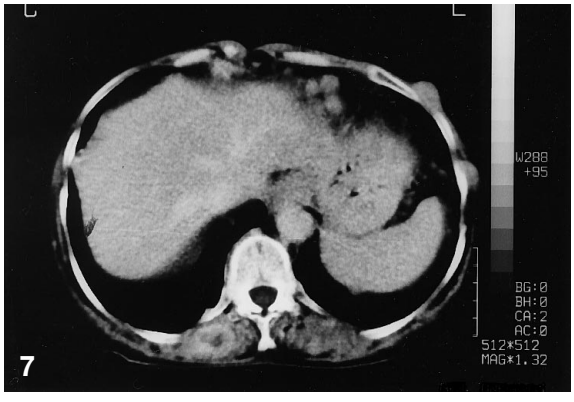
Primary LMS of the liver can arise within the liver parenchyma or the ligamentum teres. Since the ligamentum teres contains a remnant of the umbilical vein, LMS is considered to arise from the wall of this vein. Up to now, eight cases of primary LMS of the ligamentum teres have been reported [10]. To our knowledge, there is not any reported case of LMS infiltration of the ligamentum teres simultaneously with the other organs, except our case.

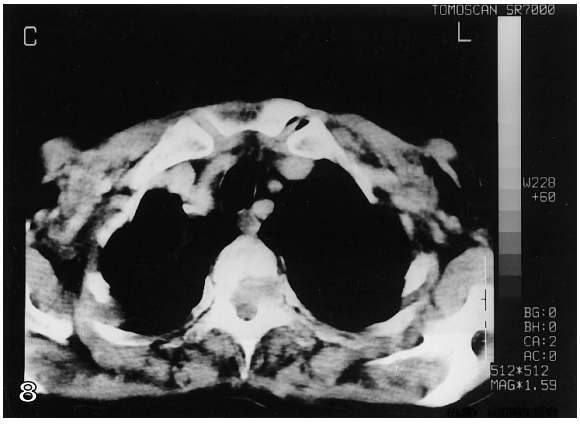
It is an uncommon feature of an extraosseously located primary LMS of the soft tissues to infiltrate the bone secondarily, since a soft tissue sarcoma tolerates natural borders, such as fasciae and bone, for a relatively long time [11]. In cases of metastasis, most of the primary tumors are localized in the retroperitoneum and in the visceral organs, especially in the uterus and in the gastrointestinal tract. In our case, we could not demonstrate any mass extending into the vertebral and iliac bones, neither the presence of a retroperitoneal nor visceral-organ LMS elsewhere, which might be considered primary. Primary LMS of the bone is an even rarer phenomenon with only 50–60 cases reported in the literature [12]. The long bones of the extremities and the bones of the oral region are the most common sites [12, 13]. Currently, the tissue of origin of these tumors is still a matter of controversy, although they are assumed to be derived from the smooth musculature of intraosseous vessels

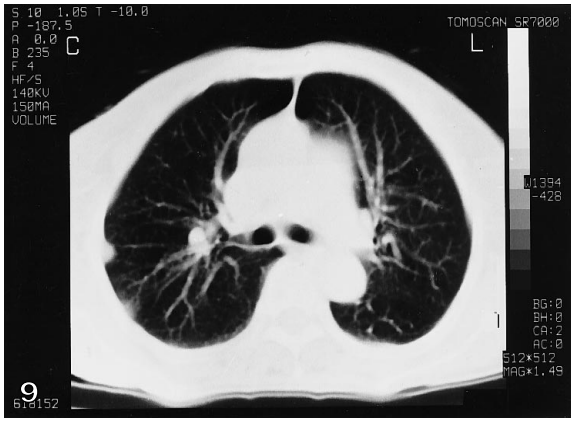












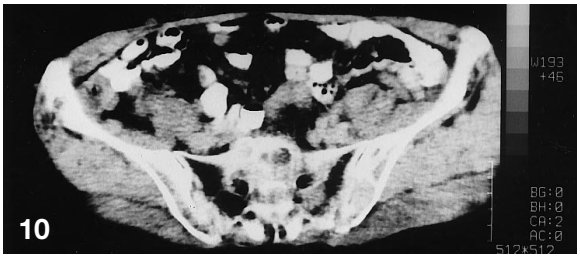


Fig. 5. **a** Ultrasound and **b** CT scan of the ligamentum teres reveal a mass *(arrow)* 15 mm in diameter. Bilateral contrast-enhanced nodular infiltration of the paravertebral muscles is also seen on CT **(b)**

Fig. 6. Ultrasound of the urinary bladder shows a hypoechogenic mass, 1–2 cm in size, indenting the anterior wall of the bladder

Fig. 7. A CT scan of the upper abdomen reveals multiple nodules involving the left intercostal muscles and right paravertebral muscles

Fig. 8. A CT scan of the upper thorax. T4 vertebra corpus is destroyed and a soft tissue mass extends into the spinal canal

Fig. 9. A CT scan of the thorax reveals multinodular involvement of the right pleura

Fig. 10. A CT scan of the pelvis. There is a moth-eaten lytic lesion at the right iliac wing

[11]. Our case is also unique for multiple deposits involving the mandibula, thoracal vertebra, and the iliac bone concurrently.

Leiomyosarcoma tend to disseminate hematogenously, and regional lymph node involvement is a rare and late phenomenon [3]. We did not observe any lymphadenopathies in any site of the body.

Although LMS is a rare phenomenon, multiple highly vascular nodular lesions small to moderate in size

should raise the suspicion for the diagnosis of this disease. The small lesions tend to be homogenously hypoechogenic on US scans and homogenously contrast-enhanced on CT scans. However, some of the lesions more than 10 mm in size show central hypoattenuation representing necrosis with peripheral contrast enhancement. Since there is no pathognomonic radiologic finding for this disease, the diagnosis can only be achieved by biopsy. However, imaging modalities provide great information in demonstrating how widespread the extent of the disease is in patients with known or suspected LMS.

In conclusion, multicentric synchronous LMS is an extremely rare pathology. Although the exact histogenesis of this tumor remains somewhat speculative, LMS may develop from any site where smooth muscle cells are located. To our knowledge, very few cases of gastrointestinal and extraintestinal LMS have been reported in the literature [14]. Considering the size and the localization of these lesions, our case is compatible with multicentric LMS.

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