Case report

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Superficial leiomyosarcoma of the scalp: a case report

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Abstract

We present the case of a 91-year old female, with no family history of malignancy, diagnosed with primary superficial leiomyosarcoma G1 of the scalp with frontal bone lysis and intracranial extension. The particularity of this case is the rarity of this tumor and the uncommon location. Also the bone involvement, present in our case, has been seen only in a reduced number of patients, approximately 10% of the cases.

Keywords: leiomyosarcoma, scalp, bone lysis

Rezumat

Prezentăm cazul unei paciente în vârsta de 91 de ani, fără istoric familial de malignitate, diagnosticată cu leiomiosarcom superficial G1 la nivelul scalpului (regiunea fronto-parietală dreaptă), cu liză osoasă frontală şi extensie intracraniană. Particularitatea cazului este raritatea acestui tip de tumoră şi localizarea sa puţin frecventă. De asemenea, afectarea osoasă, prezentă în cazul nostru, apare doar într-un procent redus, aproximativ la 10% din cazuri.

Cuvinte cheie: leiomiosarcom, scalp, osteoliză

Introduction

Sarcomas of the soft tissue are a very rare condition, comprising approximately 1% of malignant tumors [1]. Leiomyosarcomas are rare aggressive mezenchymal neoplasms, which arise from smooth muscles. The most common locations are uterus, gastrointestinal tract, and retroperitoneum. Superficial leiomyosarcomas represent only 7-10% of leiomyosarcomas cases, the most affected parts being the extremities, especially lower extremities, followed by head and neck region. The etiology is unknown; some correlations have been established with radiation and chemical exposure, trauma, chromosomal defects [2,3]. Leiomyosarcomas have been associated with bilateral hereditary retinoblastoma (result of mutations or deletions on RB 1 gene) [4].

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Case report

A 91 year old female with a prior history of cardiac disease and no family history of malignancy, was admitted for cardiac decompensation symptoms. The patient was referred to our radiology department, with the suspicion of an ischemic stroke and a left hemiplegia, with indication for a brain CT. She was disoriented and confused.

At physical examination, a scalp tumor, in the right fronto-parietal region, measuring approximately 4 cm, with normal overlying skin, without ulceration, was found (fig 1). The patient reported that the tumor had started to grow slowly one year ago and there were no symptoms during the time.

One year prior presentation a skull plain x-ray evidenced a diffuse frontal hyperostosis and the CT exam confirmed the presence of a tumor at the fronto-parietal region, slightly hyperdense and inhomogeneous, with



Fig 1. Local exam: right fronto-parietal superficial tumor.

small areas of necrosis, intracranial frontal extension and bone lysis. The tumor measured that time almost 5 cm, without displacing midline structures. After intravenous contrast, the lesion enhanced homogeneously, except the small foci of necrosis.

At presentation in our radiology department an ultrasound examination was performed- a large inhomogeneous mass, with irregular contour andtransonic areas (most likely necrosis) was found. Also small, hyperechogenic foci with the significance of calcification were present. The bone surface was interrupted and irregular, sign for bone involvement with lysis. At Doppler ultrasound examination, the tumor showed moderate vascularization (fig 2).

A new CT examination depicted the extracranial tumor, with bone involvement and intracranial extension. There was brain edema surrounding the lesion. The exact size of the tumor was 7,5x9x8 cm, with mass effect on

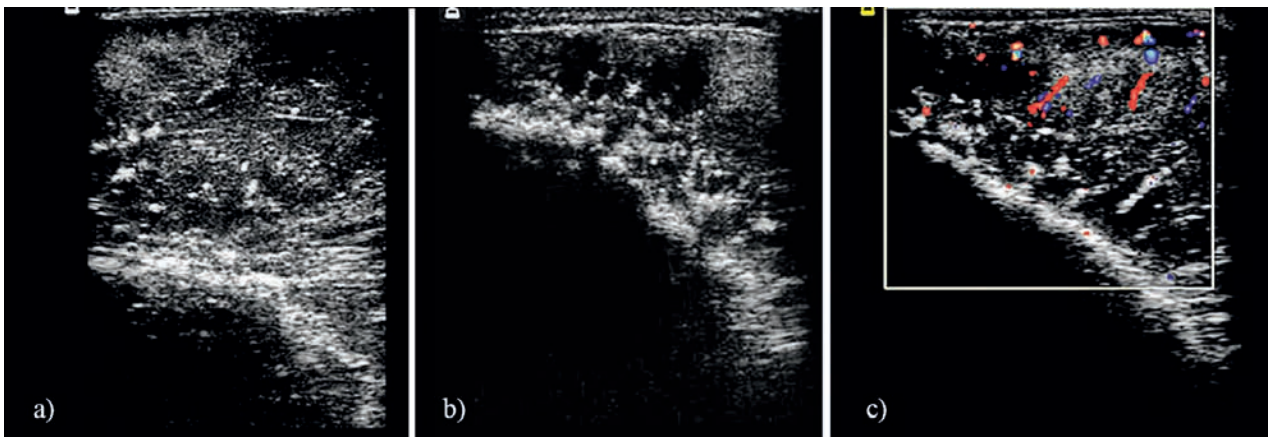


Fig 2. Ultrasonography: a) large, inhomogenous mass with necrosis, small foci of calcification and periosteal reaction; b) bone lysis; c) moderate vascularization of the tumor

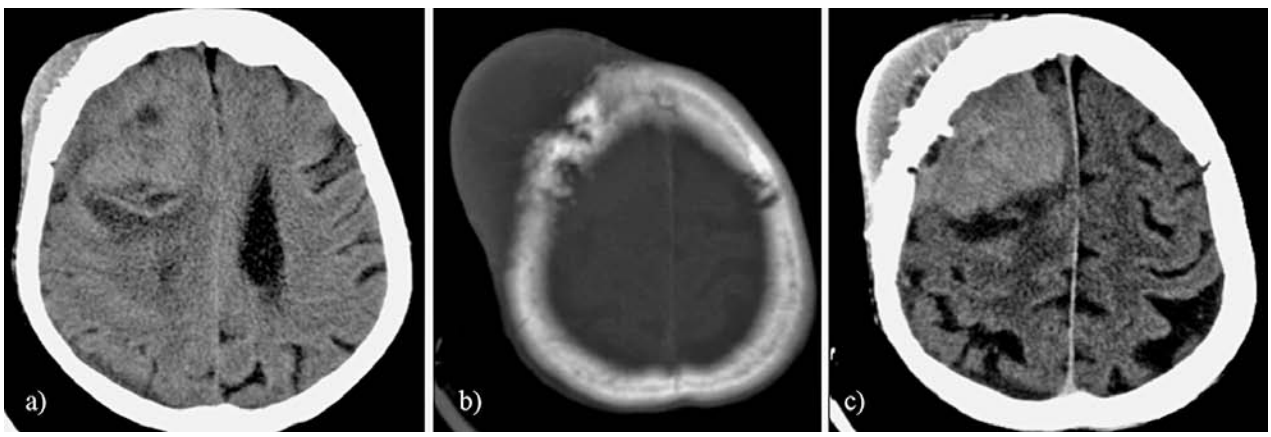


Fig 3. CT examination: a) non-enhanced CT, brain posterior fossa window: inhomogenous, slightly hyperdense tumor, with mass effect on brain parenchyma, on right lateral ventricle and middline structures b) non-enhanced CT, bone window: frontal bone lysis and periosteal reaction; c) CT with i.v. contrast, brain posterior fossa window: the lesion enhances heterogeneously.

brain parenchyma, on the right lateral ventricle and displacing the midline structures to the left with 8,5 mm. CT confirmed the inhomogeneous aspect of the tumor: non-enhanced exam showed a slightly hyperdense lesion, heterogeneous, with hypodense areas (larger than previous examination) and small foci of calcification. After intravenous contrast the tumor enhanced also heterogeneous, with the presence of hypodense, non-enhancing areas (necrosis). CT also confirmed the lysis of the frontal bone and the periosteal reaction (fig 3).

Comparing the two CT examinations, the tumor had increased in size and the brain parenchyma and intracranial structures were more affected than a year earlier. So, the final CT diagnosis based on the two exams was: an extracranial soft tissue tumor of the fronto-parietal region with intracranial frontal extension and bone lysis.

Because of the status of the patient we could not perform an MRI, but this examination, especially in superficial tumors, has shown no sensitivity or specificity higher than CT for the diagnosis or the evaluation of a tumor.

Concerning the management of this case, there was no surgical excision because of the intracranial extension at the time of diagnosis, the age and status of the patient. A biopsy was made and the histological diagnosis was: superficial leiomyosarcoma G1.

Discussion

Superficial leiomyosarcomas are subdivided in primary and secondary tumors. Usually primary leiomyosarcomas are solitary nodules, multiple nodules being more specific for secondary lesions (especially from the retroperitoneum and uterine tumors) [4]. At the head and neck region, primary lesions are present in only 3-10% of the cases. Primary leiomyosarcomas can be cutaneous (dermal) derived from arrector pili or subcutaneous type arising from the smooth muscle wall of blood vessels [5]. Any age can be affected. Some studies have shown preponderance for women, other studies proved a preponderance for men, so there is no certain prevalence for males or females [2]. Cutaneous tumors tend to be slow-growing and smaller (<2 cm) and subcutaneous type are usually larger and faster growing. Kaddu and al described two histopathologic growth patterns in cutaneuous leiomyosarcoma: nodular (the most common type) and diffuse [4].

The histological features of leiomyosarcomas are well known. The tumors are cellular and infiltrative, and most of them have a fascicular pattern of growth. The cells are moderate sized and spindle shaped. The nuclei are elongated ("cigar shaped") with blunt ends and contain perinuclear vacuoles and eosinophilic cytoplasm. Cutaneous lesions present a grey zone between them and overlying epidermis [6].

These soft tissue sarcomas are classified according to GTNM staging system, described by the American Joint Committee on Cancer. Superficial tumors are located above the superficial fascia, without invasion of the fascia; deep tumors are located beneath the superficial fascia, or invasion of the superficial fascia (retroperitoneal, mediastinal, and pelvic sarcomas are considered deep tumors). The grade is classified as: G1 – well differentiated, G2 – moderately differentiated, G3 – poorly differentiated, G4 – poorly differentiated or undifferentiated (four-tiered-system only) [7].

The metastatic potential of the two superficial forms differ: a 5-10% risk in cutaneous type and 30-40% in subcutaneous tumors. The most common site of metastasis is the lung [4]. Both types have a high recurrent rate (approximately 70%), higher on the subcutaneous tumors [5]. The survival at 5 years is 61-100%, the prognosis is better with the cutaneous form [3]. There are a few factors affecting the prognosis: high mitotic index (≥ 5mitoses per 10 high-power fields), high histological grade, extensive necrosis, nodular growth pattern, deep tumor and large size (≥5 cm) [8].

The differential diagnosis of leiomyosarcomas includes lipoma, dermatofibroma, dermatofibrosarcoma, neurofibroma, spindle-cell melanoma, spindle-cell squamos carcinoma and atypical fibroxanthoma [3].

The imaging techniques have an important role in the evaluation of the tumor. CT is useful in determining tumor extent, planning surgical therapy and assessing the presence of metastasis, especially in the lungs. MRI can provide additional information regarding neurovascular detail, but is more useful in deeply invasive lesions [4]. On MRI, in T1 sequence, leiomyiosarcomas appear as low or intermediate signal lesions, on T2 they have slightly high signal. After intravenous contrast, they enhanced intensely [1]. Ultrasound can detect the presence of the tumor, the presence of necrosis, sometimes bone involvement (irregular surface of the bone indicates bone lysis). Small tumors (less than 2 cm) tend to be homogeneous, hypoechogeneous, with vascularization present at Doppler ultrasound examination [9]. Larger tumors are inhomogeneous, with calcification and areas of necrosis. Plain x-rays are of little use in the evaluation of soft tissue sarcomas, but may evidence the modifications in the bone structure. [4].

Surgical excision is the primary treatment for superficial leiomyosarcoma. Until now, wide local excision, with 3-to 5 cm surgical margins has been recommended for both tumor types, but recent literature has demonstrated the successful treatment for superficial leiomyosarcoma

with narrow margin excision as well as with Mohs micrographic excision. Radiotherapy and/or chemotherapy have unclear benefits, appropriate adjuvant treatment, especially for high-risk advanced stages tumors remains to be determined in the future; external beam radiation is commonly administered in case of high-grade tumors [10].

In conclusion we have presented the case of a rare tumor: superficial leiomyosarcoma. The particularities of this case were the age of the patient, the uncommon localization, and the bone involvement (seen only in 10% of the cases [1]). The exactly incidence of this pathology is unknown, frequently being misdiagnosed. A review of the literature revealed differences in location between the two types of leiomyosarcoma. Subcutaneous tumors occur most frequently on the lower extremities and cutaneous leiomyosarcomas are present on the head and neck, more than any other location of the body; 48% of reported superficial cutaneous leiomyosarcomas are located on the head and neck, 21% on the trunk and 31% on the upper and lower extremities combined [10]. In 1996, Bernstein and Roenigk observed a difference in the anatomic locations of cutaneous versus subcutaneous leiomyosarcomas; the conclusion was that subcutaneous tumors were far more common located in the extremities (62%) than cutaneous tumors (33%) [11]. Recent studies proved also that the most common location for cutaneous leiomyosarcomas is the head and neck region [10]. So, even though it is a rare condition, the head and neck region, especially in elderly patients should be considered among the differential diagnosis in soft tissue tumors. A delayed diagnosis is correlated with a larger size, invasiveness into adjacent structures, with significant influence of the treatment. Surgical excision is the only and most effective way to treat this condition and avoid local recurrence [12].

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