



Case report

Surgical treatment of undifferentiated soft tissue sarcoma in a young 17-year-old woman: A case report

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ABSTRACT

This case report describes the clinical case of a 17-year-old woman with an undifferentiated soft tissue sarcoma in the left supratrochanteric area. The young woman came for observation at our plastic surgery hospital with a large vascular mass visible on her left side which also made walking difficult. Our patient reports the onset of the mass about two months earlier and its growth very quickly. In this case report, we will analyze the demolitive and reconstructive surgical procedures in order to guarantee our patient radical surgery and the possibility of continuing radiotherapy and any specific chemotherapy to avoid the risk of relapse and metastasis over time.

1. Introduction

Soft tissue sarcomas (STMs) are tumors that arise in the muscles, tendons, synoviums, adipose tissue and connective tissues in general. Adult STMs are rare cancers. Their overall incidence is around 3–5 cases/100,000 inhabitants/year. They therefore represent 1 % of adult cancers. In the USA there are approximately 8700 cases/year, while in Italy an absolute number of 2300 cases/year is estimated. They become even more rare if they are broken down in the various histotypes. The incidence trend is stable over time, albeit with a modest upward trend in women. The trend of the incidence in relation to age shows a first peak in pediatric age then a plateau and subsequently a tendency to increase the incidence rather constant with the starting age from the age of 20 with a maximum peak after the age of 60. More than 50 types are grouped under the term of STM different histologicals with specific biological characterizations and distinct behaviors [1]. Most STMs are sporadic. Few have an identifiable cause. There is an association between Epstein Barr Virus viral infections in patients with AIDS and leiomyosarcoma. Sarcomas secondary to radiation are evident after treatment for lymphoma, carcinoma of the portio or breast cancers.

Unfortunately, a diagnostic delay is still frequently found, partly connected with the rarity of pathology, which leads to underestimation of soft tissue masses and misdiagnosis. Taking into consideration that an optimal surgery is all the more feasible the smaller the volume of the sarcoma, it is considered appropriate to raise awareness among General Practitioners and Outpatient Specialists on the existence of STMs to avoid diagnostic delays and increase the number of timely diagnoses [2,3]. In this paper, we explored a case of a 17-year-old woman with an undifferentiated spindle cell soft tissue sarcoma in the left supratrochanteric area. This work has been reported in line with the PROCESS criteria [4].

2. Presentation of case

In March 2022 we visited a 17-year-old woman in the plastic surgery department of the San Carlo Hospital in Potenza (Basilicata, Italy). The patient presented a large and visible mass in the left supratrochanteric area. The patient reported that this mass had grown rapidly in less than two months and prevented her from walking and leading a normal life. The mass had a very macroscopically vascularized appearance, with a

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visible venous reticulum and an almost necrotic appearance at the apex of the mass. The patient reported that she had no pain, only a sense of external pressure in the left side which became more important every day. The patient's blood tests were all normal. The patient has no noteworthy medical history, no concomitant pathology, no history of drug or alcohol intake. The family history does not reveal any oncological pathologies. There is no relevant genetic information. MRI was conducted in multiple planes, employing GRE and FSE T1 and T2 frequencies weighted with and without suppression for adipose tissue and supplemented with T1-weighted scans after contrast. The examination documents, in correspondence with the upper area of the left thigh, the presence of a solid nodular lesion of 10 × 10 cm extra-fascial with the presence of a necrotic/haemorrhagic hypointense central core. The lesion has irregular margins without a clear plane of cleavage with the ipsilateral gluteus muscle. The mass described is highly vascularized and has the superior gluteal artery as its main vascular afferent. The examination reveals no apparent signs of bone infiltration. We subsequently performed an incisional biopsy examination for the typing of the lesion. A high-grade, undifferentiated soft tissue sarcoma was found on biopsy examination. Before surgery, we performed the staging of the disease through global body tomoscintigraphy (PET-TC). The examination revealed a large area of dense and pathological accumulation of 18F-FDG, characterized by central photopenia, as well as by colliquative necrosis in the left supratrochanteric area. No further pathological findings were found in other areas of the body of 18F-FDG hyperaccumulation, thus indicating that the disease has not metastasized for now (Fig. 1). At this point we immediately carried out the surgical treatment trying to be as radical as possible.

The surgical excision was carried out with wide margins to make the surgical act more radical. The surgical procedure was carried out by the Plastic Surgery team of the San Carlo Hospital in Potenza. We can see the preoperative state of the patient, the lesion sent for histological examination and the area reconstructed with the use of a double layer dermal matrix (INTEGRA) and autologous skin graft (Fig. 2).

The histological examination of the lesion gave important information for the continuation of oncological therapies. On histological examination, the lesion appears to consist of a proliferation of predominantly spindle cells, organized in bundles with large areas of necrosis, sparing the lateral and deep surgical margins. High mitotic index neoplastic cells expressed the following immunophenotype: vimentin+, CK AE1\AE3-, EMA-, CD34-, NSE-, actin, desmin-, S100-, MDM2-, melan A-, Cd68, CD99 not evaluable, KI 67: 25 %. Such immuno-morphological aspects lead to undifferentiated spindle cell sarcoma worthy of genetic and biomolecular investigations. At this point the patient was followed up at an oncological level and underwent cycles of radiotherapy in order to minimize the risk of localized relapse and distant metastasis. The patient was also sent to specific genetic and biochemical studies.

3. Discussion

Surgical treatment is indicated in patients with sarcomas of the limbs and trunk. When the disease is local, the first instance treatment is surgery. This can be integrated with radiotherapy and chemotherapy. Surgery (\pm radiotherapy) aims at the local control of the disease which today is reached in 90 % of cases at 5 years. Surgery must have adequate margins, trying to reduce functional damage and aesthetic. Surgical margins are defined as adequate when they are radical or wide; not adequate when they are marginal or intralesional. By radical margin we mean the resection of the entire anatomical compartment where the tumor is located; sometimes to obtain this margin it is necessary to amputate the limb. For this reason an intervention radical often presents disabling functional sequelae, and is generally replaced by an intervention large, integrated with pre or postoperative radiotherapy. In many cases, however, the sarcoma does not arise in defined anatomical compartments and therefore the resection radical or compartmental is not possible. In these cases it is necessary to obtain a large surgical margin (generally where possible >1 cm). The intervention is defined as large when the margins are made up of healthy tissue in all directions or, in proximity to non-expendable critical structures, from quality tissue such as the periosteum, the perineurium, the adventitia of the vessels, the muscle bands, of any thickness as long as microscopically negative. If the sarcoma is high grade and/or deep seated and/or >5 cm in size, surgery should be completed by local radiotherapy. Extensive surgery associated with radiotherapy results in local control similar to radical resection. Radiotherapy is never a remedy for inadequate surgery, but must be used to complete an oncologically correct surgery. Inadequate margins are the marginal margin (removal in blockage of the tumor by passing through its peripheral reactive pseudocapsule) which it can leave in situ neoplastic and satellite digitations and the intralesional margin (excision performed through the mass tumor) that leaves macroscopic parts of the tumor in situ. A marginal margin must be considered acceptable only if it cannot be improved, if not at the cost of serious damage functional or aesthetic. For this reason, focal surgical marginality can be accepted only in contiguity with critical structures (prominent vessels or nerves). When a marginal resection is foreseeable it must preoperative radiotherapy should be considered to favor the lower risk of marginalization and relapse. A marginal surgery must always be followed by radiotherapy, in the intermediate and high degree forms, while it can be considered sufficient in some low-grade forms when a possible one is foreseeable conservative surgical recovery (\pm radiotherapy) in case of relapse (i.e. well differentiated liposarcoma of the limbs, dermatofibrosarcoma protuberans especially if affecting the head and neck). An intralesional intervention or an unplanned marginal intervention cannot be cured with radiotherapy e it must absolutely be radicalized surgically [5,6].

Radiotherapy, whose purpose is the control of local disease, can be performed in the preoperative or postoperative. Radiotherapy can never heal inadequate surgery. The optimal mode of association between

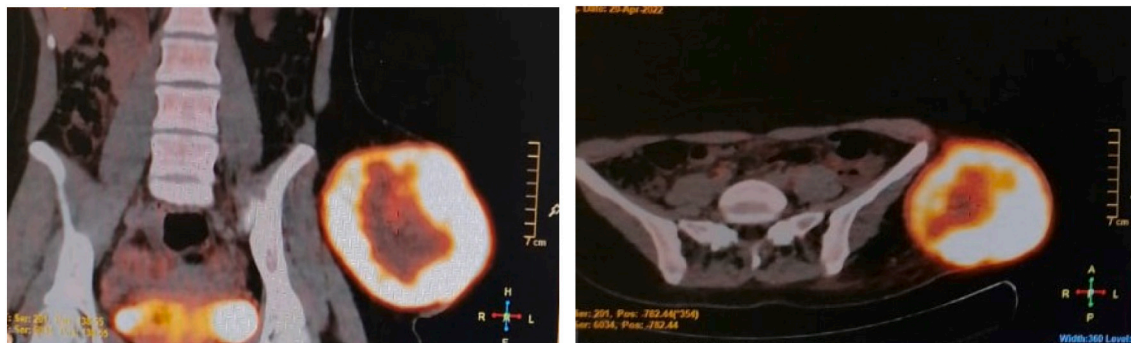


Fig. 1. Hyperaccumulation of 18F-FDG at the level of the mass in the left supratrochanteric area on PET-CT.

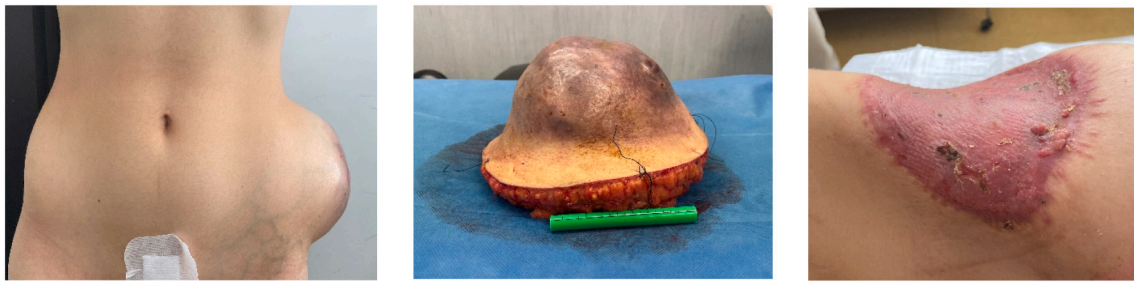


Fig. 2. In order preoperative image followed by the mass removed and sent for final histological examination and finally image after reconstruction with artificial dermis and autologous skin graft.

surgery and radiotherapy is not yet defined. Indeed it is not demonstrated by randomized studies which of the two approaches is the best. The choice must be made after multidisciplinary discussion of the case. The postoperative radiotherapy completes the large surgery or the marginal surgery that cannot be improved intermediate or high grade sarcomas, especially if larger than 5 cm in diameter or located in deep sites. It can also be proposed in low-grade, large-volume sarcomas, especially if they occur in sites critical or profound, or with non-radicalizable margins. The goals of concomitant or sequential preoperative treatment include: (a) to reduce the risk of local recurrence in tumors that are marginally resectable or at high risk of recurrence, (b) to reduce the risk of distant recurrence, (c) reduce the total radiation dose to minimize post-actinic complications, (d) provide prognostic information based on the response to treatment and guide any choices postoperative therapeutics. In the neoadjuvant setting, an integrated radio-chemotherapy treatment can be considered especially in patients with high-grade, locally advanced sarcomas localized to the limbs. They were evaluated both the sequential approach (CT followed by RT) and the concomitant approach (RT/CT). Although in both cases there are promising results in terms of preservation of functionality and reduction of recurrence local, with no adverse effects on survival, currently available data do not allow to consider this approach is a standard of care. The integration of chemotherapy with radiotherapy in the post-operative period can be considered in cases selected: it is reasonably feasible especially in patients with sarcomas at high risk of local or distant recurrence (>5 cm, high grade and deep), especially if localized to the limbs and superficial trunk [7–16]. The patient underwent adjuvant radiotherapy. The dose administered was 50 Gray on the left gluteal region, plus a concomitant overdose of 61.6 Gray for a total of 28 sessions performed over a few months, using 6 V megavolt IMRT photon beams.

4. Conclusion

Our clinical case is undoubtedly an exceptional event, considering that our patient is only 17 years old. The incidence of developing high grade undifferentiated spindle cell soft tissue sarcoma in the young population is extremely low. In this case it was extremely important to have carried out radical surgery, with negative resection margins, which allowed radiotherapy to be carried out in the immediate post-operative period. Furthermore, our patient did not show distant metastases on PET-CT with a much better long-term prognosis, especially after radiotherapy and possible chemotherapy. Genetic and biomolecular tests will be extremely important for this young woman, in order to be able to act on specific targets for cancer therapy.

Consent

Written informed consent was obtained from the patients for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

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Ethical approval

Exemption for this study has been given by our institution.

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Guarantor

Federica Giuzio, MD, PhD.

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1. Name of the registry: -
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CRediT authorship contribution statement

Giuzio Federica: Conceptualization, surgeon. Writing - Original draft preparation.
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 Luigi Mele: Visualization, Investigation.
 Brongo Sergio: Supervision.
 Saturnino Carmela: Validation.
 Giuliani Antonio: Writing - Reviewing and editing.

Declaration of competing interest

None.

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