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Unusual bleeding of a giant cell fibroblastoma: a soft tissue sarcoma of the skin mimicking metastatic melanoma

A 56 year-old man presented to the emergency department after a spontaneous bleeding of a giant mass located on the right axilla. Clinical diagnosis was recurrent hemorrhagic nodular melanoma. Ten months previously a malignant melanoma had been removed from the dorsum by radical excision and surgical margins had been disease-free (MM: Breslow IV, Clark IV, lung and lynphnode metastases).
The patient required immediate emergency surgical intervention to prevent death by hemorrhagic shock. The tumor was bleeding and the patient required a transfusion. Subjective symptoms included pain in palpation and spontaneous hemorrhage, poor general appearance, pale skin, BP 80/40 mmHg, HR 100/min with overall symptoms of hypovolemic shock. At the time of surgery, radical tumor excision was performed with an approximately 3 cm circumferential gross tumor free margin. The resultant defect was reconstructed by pectoral rotation fascio-cutaneous flap. The histological diagnosis demonstrated an undifferentiated high-grade pleomorphic sarcoma with microscopic tumor free margins.

KEY WORDS: Melanoma, Skin bleeding, Soft tissue sarcoma

Introduction

Soft tissue sarcomas (STS) are a heterogeneous group of rare solid tumors with distinct clinical and pathological features. Multimodality treatment (surgery with radiotherapy and or chemotherapy) is often used for the management of the patient with intra-abdominal or retroperitoneal localization. Sarcomas constitute less than 1% of all cancer. Radiation therapy seems to be a risk factor; STS often originate in radiation fields following therapeutic irradiation for a variety of malignancies. Also chemotherapy seems have a correlation with this kind of tumor, the drugs implicated include cyclophoshamide, melphalan, procarbazine and other chemiotherapic agent and the relative risk of sarcoma appears to be increased by cumulative drugs exposure.
As well known melanoma is a type of skin cancer which forms from melanocytes and it is the most malignant tumor of the skin. In women, the most common site is the legs, and melanomas in men are most common on

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the back. It is particularly common among Caucasians, especially northern and northwestern Europeans, living in sunny exposure. There are higher rates in Oceania, North America, Europe, Southern Africa, and Latin America. This tumor originate from the pigment-producing melanocytes in the basal layer of the epidermis when unrepaired DNA (often caused by ultraviolet radiation of the sun) trigger mutation that lead the skin cell multiply rapidly and form tumors.
Often STS look like a secondary localization of melanoma because it shows at histology brown granular pigmentation that can be identified like melanin [1,2].
Because melanoma has often cyto-morphologic and architectural charateristics overlapping with many mesenchymal neoplasms, especially in its amelanotic variant, to distinguish the two it is necessary an immune histochemical study by means of antibodies (Melan A, PNL2 S100 and vimentin).

Case Report

A 56 year-old male patient was admitted to Emergency Department for the bleeding of an ulceration localized on a huge right axilla tumor. The patient required immediate emergency surgical intervention to prevent his death by hemorrhagic shock.
The first episode of bleeding had occurred one month earlier and stopped spontaneously.
Physical examination showed a large, bilobed tense mass measuring 200 × 135 × 122 mm, located between the posterior axillary line and the right pectoral region (Fig. 1), infiltrating skin and the deeper soft tissue layers of the right axilla.
The tumor appeared painful at palpation, ulcerated, phlogistic, hemorrhagic, fast growing, dark-brownish and presenting in its central part an ulceration with active bleeding.

The initial clinical diagnosis was recurrent nodular melanoma because ten months previously a malignant melanoma had been removed from his dorsum by radical excision and surgical margins had been demonstrated disease-free (MM: Breslow IV, Clark IV, lung and lynphnode metastases absent). The patient showed poor general appearance, pale skin, BP and HR not valuable and mainly symptoms of hypovolemic shock.
Laboratory tests showed anemia (hemoglobin 6 g/dl, RBC 2.70 10^6/mm^3) and leukocytosis (WBC 16 10^3/mm^3). Chemistry were as follows: total protein 5.5 g/dl, albumin 3.1 g/dl, blood urea nitrogen 35 mg/dl, creatinine 1.43 mg/dl (indicating dehydration), total bilirubin 1,33 mg/dl, aspartate aminotransferase 11 U/l, alanine aminotransferase 9 U/l, creatinine kinase 18 U/l, Tomodensitometry revealed a voluminous solid mass (transverse diameter 20 cm) formed by very vascularized tissue delimiting colliquate areas that didn't infiltrate the vascular axis of the axilla.

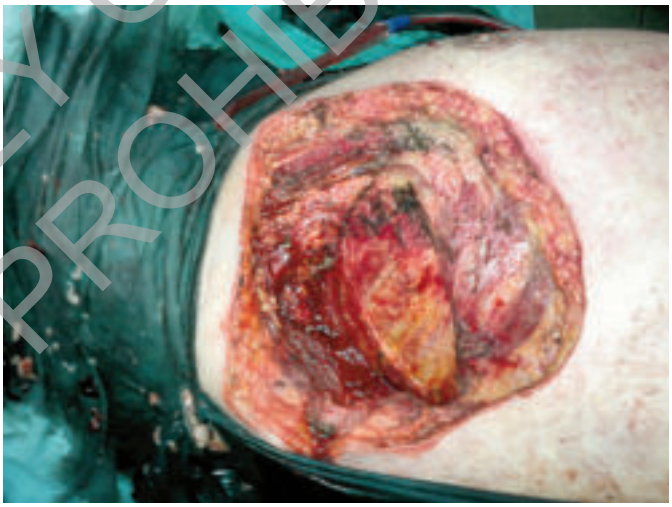


Fig. 2

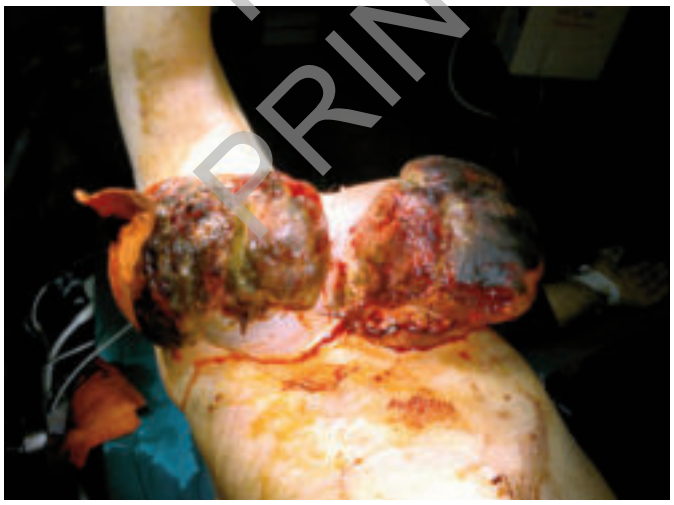


Fig. 1

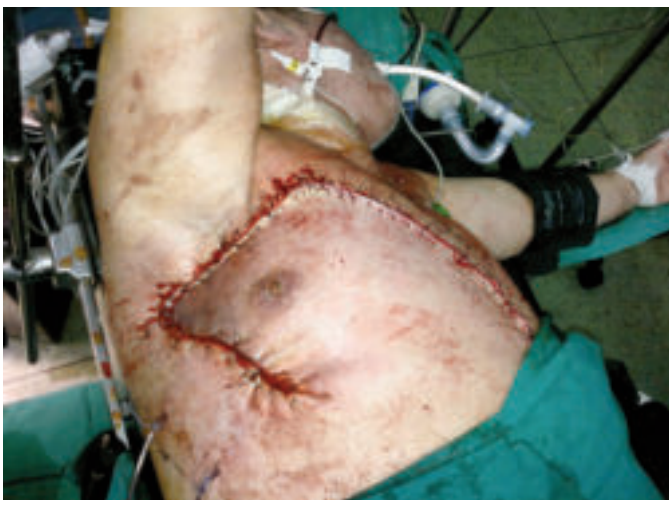


Fig. 3

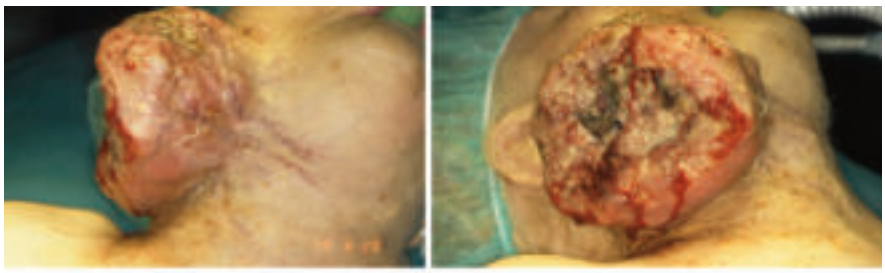
All attempts to stop the bleeding failed and after applying a compression bandage and transfusion of 3 units of erythrocytes, the patient was addressed to an emergency surgical intervention.
At the time of surgery radical tumor excision was performed with an approximately 3 cm circumferential gross tumor free margin. Tumor resection was enlarged to latissimus dorsi, serratus and pectoral muscles excision (Fig. 2).
The resultant defect measured approximately 15 cm by 20 cm, and the gap was covered by pectoral rotation fascio-cutaneous flap (Fig. 3).
The patient was discharged on the 10th day in good general condition and left to the oncological care.
First intention wound healing without complications was noted. Sutures were removed on day 15.
The patient's post-operative course was uncomplicated. No adjuvant radio- or chemotherapy was administered. Full functional and almost full cosmetic recovery was achieved; follow-up for more than two following months has been uneventful.
Excision biopsy of the current lesion revealed an infiltrative, well-vascularized mesenchymal neoplasm containing pleomorphic tumor cells, increased mitotic count with some atypical mitoses. Immunohistochemistry was strongly positive for desmin, miogenine, S-100 in condroid areas. Positive areas for CD99, negative Pancytokeratin, Melan A, HMB45.
The histological analysis revealed a "sarcoma" contains pleomorphic tumor cells with condrosarcomatoid and a rabdomiosarcomatoid area, and tumor-free surgical margins up to 3 cm.

Discussion

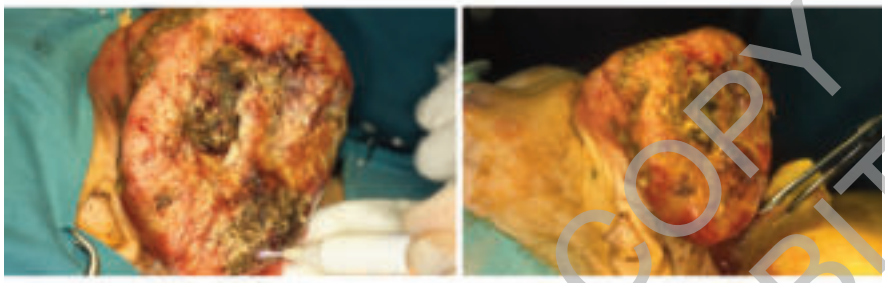
Sarcoma is a biphasic cancer comprising malignant heterologous elements of both epithelial and mesenchymal cells. It has been described in other organs including the uterus, ovary, lung, bladder, breast and larynx. Primary cutaneous case is rare and only few cases are reported in literature.
Soft-tissue sarcomas (STS) are a relatively rare disease accounting for approximately 1% of adult malignancies. The coexistence of STS and melanoma in the same individual, either synchronous or metachronous has been reported in a few studies throughout the world, prospecting the possibility of an association between the two [2]. Some workers have even hinted at a "melanoma/sarcoma syndrome" [4,5]. Somatic mutations in CDK4, MDM2 or p53 genes have been proposed as a common pathogenetic pathway for these two tumors [5,7].
Several investigators proposed an association between melanoma and sarcoma has been proposed [8,9]. Berking and Brady [4] reported 48 patients with melanoma and sarcoma. Among them, five patients (10%) presented with both tumors con- currently; 34 of 43 (79%) were diagnosed with melanoma first. The median interval between the two diagnoses was six years.
Twenty-five percent of the patients had additional primary malignancies, and 50% of the patients had positive family histories of cancer, suggesting a predisposition for cancer in these family teams [6].
Significant and refractory bleeding from soft tissue sarcomas appears however to be a rare event [9]. A review of the literature (a PubMed search from 1990 to 2014 with index words: cutaneous sarcoma, skin sarcoma, emergency treatment, resection, bleeding) failed to identify any reports of tumor resections performed for refractory bleeding.
Skin cancer surgery for acute indications is a rarity. The available literature describes no cases of patients with skin cancer of similar type and location, or operated on for acute indications such as bleeding. In the presented case cancer was diagnosed only after surgical excision for vital indications because of hemorrhage. In the described case it had been growing for 1 years and showed local malignancy infiltrating adjacent tissues, including blood vessels, causing their destruction and hemorrhage threatening the patient's life.
Sarcoma's primary therapy is predicated on surgical resection with an adequate margin of normal tissue [10].
Treatment of soft tissue sarcomas requires an individual plan, which considers interdisciplinary recommendations and the various clinical situations. Anatomical region, histological grading and typing, and tumor size necessitate adjusted methods of reconstruction. In general surgical resection of the tumor will be the treatment of choice [11].
Reconstructive surgery facilitates treatment of patients with soft tissue sarcoma by permitting tumor resection with adequate margins, protects vital structures, enables early postoperative radiation therapy and if necessary assists in palliative procedures. The ability to maintain function and aesthetics after tumor resection, and effective palliation improves the quality of life for these patients [12]. Tumor excision with surgical margin proved to be not only a life-saving procedure (hemorrhage control) but also a radical oncologic treatment, although it is too early to draw any final conclusions due to the short patient observation period. In conclusion, surgical excision of bleeding sarcoma with surgical margin can have a good early therapeutic effect. Although seemingly radical in the face of incurable disease, surgery can allow an extended or improved quality of life.

Riassunto

Gli autori riportano il caso di un importante sanguinamento in un paziente di sesso maschile dell'età di 56 con una voluminosa neoformazione cutanea tra la regione ascellare e pettorale di destra. La presenza di una anamnesi positiva per escissione di melanoma cutaneo avanzato avvenuta otto mesi prima aveva fatto orientare



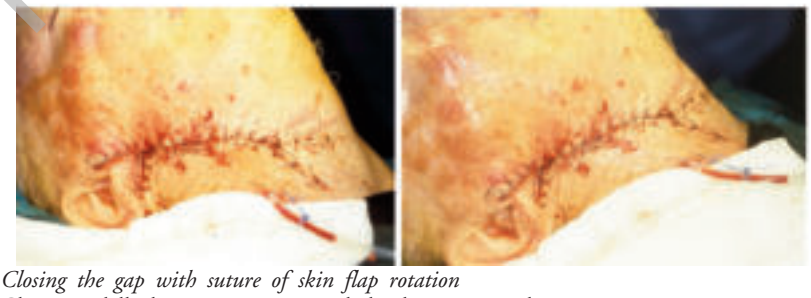
Hemorrhagic mass bordering at presentation the parotid region and the right lateral cervical one
Presentazione della massa emorragica al confine con la regione parotidea e la cervicale laterale destra



Effectiveness of preliminary temporary surface hemostasis surface with Argon Beam coagulator
Efficacia dell'emostasi preliminare temporanea con Argon Beam coagulator

En-bloc detachment of the neoplastic bulk from underlining normal tissues in a bloodless field
Distacco in blocco della massa neoplastica dai tessuti profondi in un campo esangue

Recognition and preservation of duct Stenon and branches of the facial nerve
Riconoscimento e conservazione del dotto di Stenon e di rami del nervo facciale



Closing the gap with suture of skin flap rotation
Chiusura della breccia con sutura di lembo cutaneo di rotazione

inizialmente per una mestasi cutanea da melanoma. Invece l'esame istopatologico effettuato in seguito all'escissione chirurgica avvenuta in urgenza per sanguinamento attivo non controllabile con i comuni mezzi di emostasi ha dato luogo al riscontro di un voluminoso sarcoma bilobato dei tessuti molli. La continuità cutanea è stata ristabilita mediante lembo muscolare di rotazione. Il paziente è stato dimesso il decima giornata postoperatoria in via di guarigione ed affidato agli oncologi. Una accurata revisione della letteratura scientifica mostra come la contemporanea presenza di un melanoma cutaneo e di un sarcoma dei tessuti molli attivamente sanguinante sia un evenienza molto rara. Il trattamento chirurgico demolitivo resosi necessario per il controllo dell'emostasi si è mostrato essere efficace e ben tollerato da parte del paziente.

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Commento e Commentary

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Sebbene rare, le esperienze riguardanti la necessità di escissione di masse neoplastiche sanguinanti, anche semplicemente per motivazioni emostatiche, non sono tuttavia del tutto eccezionali, e pongono talvolta problemi per coniugare esigenze demolitive con quelle conservative delle strutture nobili, pur senza fare molto affidamento sulla possibilità di miglioramento prognostico a lungo termine.
Qui è un esempio di un caso personale non pubblicato riguardante una voluminosa massa sanguinante alla base del collo di una anziana donna, anemica per lo stillicidio ematico infrenabile che bagnava rapidamente il cuscino, sottoposta di necessità a exeresi chirurgica immediate previa emostasi superficiale con l'uso dell'Argon beam coagulator. La preparazione anatomica finale, su campo operatorio esangue, consentì l'individuazione e la conservazione del tronco del nervo facciale e del dotto di Stenon.

* * *

Although rare, the experiences regarding the need for excision of bleeding neoplastic masses, simply for hemostatic purpose, are not however completely exceptional, and sometimes they pose problems if the needs of demolition should be combined with the ones conservative of nobler structures, without entrusting the chance of positive prognostic results in the long term.
Here is a not published personal case, example of a voluminous bleeding mass at the base of the neck of an elderly woman, anemic for uncontrollable oozing that bathed quickly the pillows, necessarily subjected to surgical excision after immediate surface hemostasis with the use of Argon beam coagulator. The final anatomical preparation, on a bloodless field, allowed the identification and preservation of the trunk of the facial nerve and of the Stenon's duct.
As the case presented the pathology demonstrated a sarcoma.