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Case Report

The medico-legal observation of an aggressive urogenital fibromatosis with isolated development not related to any traumatic event

Enrico Muccino [a], Guendalina Gentile [a], Stefano Mantero [b], Matteo Marchesi [c], Alessandra Rancati [a], Riccardo Zoja [a,*]

[a] Sezione di Medicina Legale e delle Assicurazioni-Dipartimento di Scienze Biomediche per la Salute- Università degli Studi di Milano, Via Luigi Mangiagalli 37, 20133 Milano MI, Italy
[b] Centro Nazionale delle Ricerche–Istituto di Ricerca Genetica e Biomedica (IRGB)- Istituto Clinico Humanitas, Via Manzoni 113, 20089 Rozzano MI, Italy
[c] Azienda Ospedaliera Papa Giovanni XXIII-Piazza OMS 1, 24127 Bergamo

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ABSTRACT

Desmoid tumor is a fibroproliferative neoplasm with an intermediate malignancy and it can be localized in every bodily district: some locations are considered exceptional, like the urogenital localization. The Author point out a rare case of giant idiopathic scrotal fibromatosis that was found during an autopsy.

A widower, that lived alone in poor hygienic conditions, was found dead in his house. The Judicial Authority ordered the autopsy, that was performed two days later at the Medico-Legal Section of Milan University.

External examinations revealed only the considerable dimension of the scrotum (cm 24 × 41). The cause of death was fixed in a cardiac tamponade due to a natural heart laceration localized in correspondence of a transmural infarction. The toxicological exam resulted negative, while the histopathological and immunohistochemical analysis qualify the scrotal mass as a desmoids tumor.

Due to the absence of predisposing conditions and of fibroproliferative infiltration in bladder and retroperitoneal space, the neoplasm was configured as an idiopathic desmoid tumor. The presented case gives the reason for the discussion concerning medico-legal aspects that are typical of rare neoplasms.

1. Introduction

The aggressive fibromatosis or desmoid tumor is a rare [1,2] and usually solitary monoclonal [3] fibroproliferative neoplasm [4,5] of the connective tissue; it sometimes can be presented in multifocal lesions [6]. It is a tumor with a slow growth [7] and an intermediate degree of malignancy (locally aggressive) [8], because it ranges from benign forms to malignant forms like sarcoma [9,10] and fibrosarcoma [11]. The differential diagnosis from malignants forms [12] is set up by the absence of mitotic anomalies, nuclear atypia [13–15] and giant cells – in the specific literature it is described only a case of transformation of a desmoids tumor in a fibrosarcoma, due to radiant therapy [16] -. According to the recent WHO classification of February 2013, the desmoids tumor is included in the group of non-metastatic and locally aggressive mesenchymal neoplasms, and it represents the 0.03% of all tumors and the 3% of soft tissue tumors [17].

According to the localization [18], desmoid tumors are divided in abdominal (more frequent) [19] and extra-abdominal, that are rarer [18]. The former involve (in about 50% of cases) [20] the anterior internal oblique muscle and the abdominal rectus muscle [21,22] and more rarely, in the 8% of cases [23], (with a different biological behavior) [24] the interior of the abdominal cavity [25], localizing at the mesentery, the retroperitoneum and the pelvic cavity [15,26]. The extra-abdominal presentation is infrequent and implicates the head (basicranium [27], paranasal sinuses [28], cheeks [29], upper lip [30] and submandibular region [31,32]), the neck [30–33] (sternocleidomastoid muscle [12,34], larynx [35]), the thorax [36] (8–10%) [37] (breast [38], ribs [39]), upper and lower limb (scapula [40,41], arms [42], hands [43], gluteus [5], fibula [44], tibia [45] and feet [46]). The localization at the urogenital tract is considered exceptional [4,47–50]: literature reports only few cases, concerning (in the man) the spermatic cord [15,50], the scrotum [51] the parascrotal region [47] and (in the woman) the vulva [52] and the ovaries [53].

The Authors report a case of death with an unexpected detection of a giant scrotal neoformation; the histopathological and immunohistochemical analysis allow to classify the neoplasm

* Corresponding author. Tel.: +39 02 5031 5685; fax: +39 02 5031 5724.
E-mail address: riccardo.zoia@unimi.it (R. Zoja).

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as an aggressive fibromatosis or desmoids tumor, with an extra-abdominal localization and a benign course. The rarity of the localization and the huge dimensions of the tumor mass constitute the motivation for the reporting of the case, that is exceptional in a medico-legal context. This article highlights the medico-legal aspects emerging from the diagnosis of rare tumours.

2. The case

A 67-years old widower was found dead in the bedroom of his house by some relatives; the house was in poor hygienic condition. The Public Prosecutor ordered the autopsy, that was performed two days later at the Medico-legal unit of Milan University. The relatives were summoned for the official cadaver identification, and they referred that the man lived alone (after the death of his wife) and refused the contacts with everyone. He suffered from an ischemic cardiomyopathy and he walked with difficulty due to an enormous inguinal hernia that was not subjected to a medical examination.

3. Results

3.1. Autopsy

The cadaver of a white Caucasoid man was in good condition of preservation, with a weight of 89 kg and a height of 172 cm, but it was in bad hygienic conditions. The external examination revealed decubitus ulcers localized at the sacral-coccygeal region and an abnormal scrotal sack, with the skin presenting multiple dyschromic and thickened areas together with deep, irregular fissures and necrotic areas. At the base of the left gluteus there was a hard, reddish and nodulated neoformation (Fig. 1).

At the thorax dissection, the pericardium sack was replete with about 700 cc of blood (with a clot of about 350 cc). The heart was enlarged (with a weight of 650 g); it also presented, in correspondence of the basis of the posterior wall, a large epicardic circular bruise (about 3.0 cm) with an irregular and blood-infiltrated laceration of about 0.4 cm. The correspondent myocardium was interested by a large transmural breach, with irregular and blood-infiltrated edges, sized about 4.5 × 2.4 cm; the surrounding area was characterized by pale muscle, with decreased compactness. All the coronaries showed thickened walls, with eccentric atherosclerotic plaques determining the decrease of the lumen size approximately of 2/3 (left coronary) and 1/3 (right coronary).

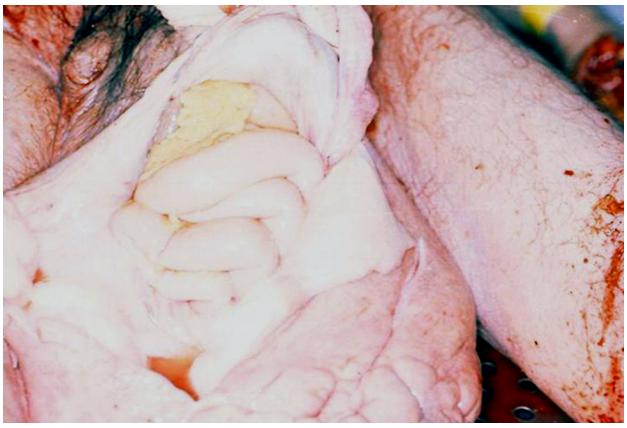

Fig. 2. The dissection of the scrotal sack: in the superior part there is the herniation of small intestine, while in the inferior part there is the lardaceous mass with soft compactness.

The abdominal dissection revealed that the colon had undamaged and normotrophic walls, but it was dislocated in the left lower quadrant, similarly to the small intestine, which also had undamaged walls: a segment of the small intestine (that was far from the Treitz about 150,0 cm) was equally dislocated in the left lower quadrant. The retroperitoneal space was normal. Concerning the urogenital tract, the kidneys, the ureters and the bladder were undamaged; the scrotal sack showed enlarged dimensions (longitudinal axis: 41.0 cm; transverse axis: 24.0 cm) and it presented normal compactness at the superior portion, while it was grayish with a "brainy" aspect and a harder compactness at the inferior portion. The serous membrane was smooth and normal; the inguinal channel, abnormally dilated, was occupied in his superior portion by a herniation of small intestine (a tract of jejunum and ileum), while the inferior portion was constituted by a grayish mass of 14.0 cm with soft compactness (Fig. 2).

At the end of the autopsy, the cause of death was identified in a cardiac tamponade originated by a natural heat breakage that was localized in correspondence of a transmural myocardial infarction. Multiple samples of biological liquid and internal organs were collected, in order to perform toxicological and histopathological analysis.

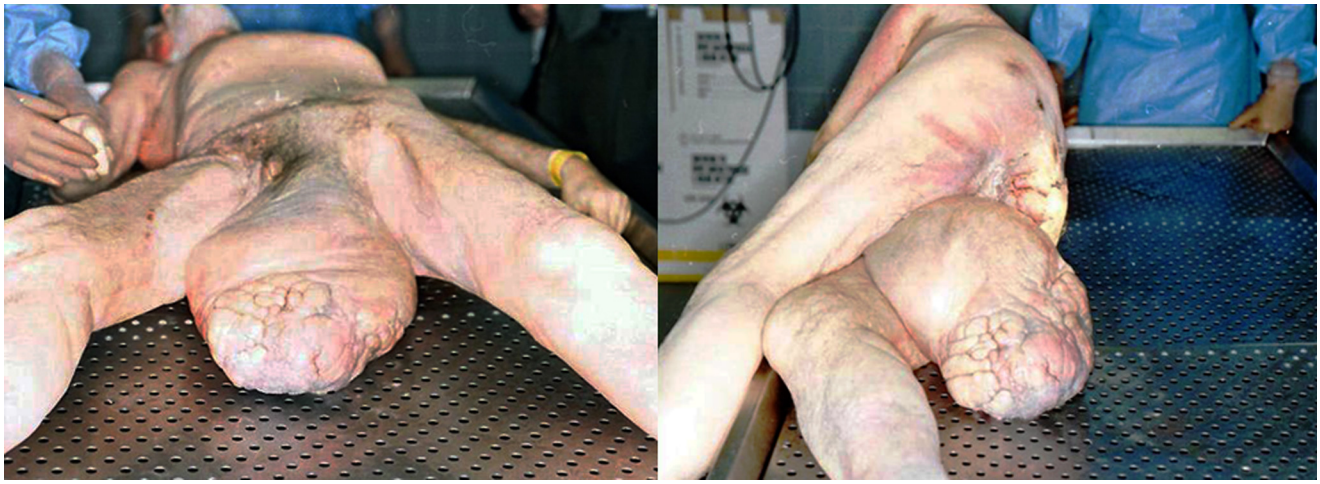

Fig. 1. The appearance of the sacro-coccygeal region and of the scrotal sack, showing different structures of the upper and inferior part of scrotal sack.