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Case report

Primary angiosarcoma of the breast complicated by the syndrome of disseminated intravascular coagulation (DIC): Case report and literature review

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ABSTRACT

Primary angiosarcoma of the breast (PAB) accounts for 0.04% of all breast malignant tumors. It affects young women usually at third or fourth decades of life. PAB clinically manifests as a painless, movable mass with sharp limits. A bluish red discoloration of the overlying skin is often observed. Enlargement of axillary lymph nodes generally does not occur.

Angiosarcoma of the breast has a very poor prognosis due to the tendency to metastasize haematogenously and high frequency of local recurrence.

Mastectomy and chemotherapy are preferable treatment choices.

This paper presents a case of primary angiosarcoma of the breast with a syndrome of disseminated intravascular coagulation (DIC).

1. Background

Primary angiosarcoma of the breast (PAB) is a rare and extremely aggressive neoplasm of the endovascular origin. Two hundred and nineteen cases have been documented[1] since the first case reported by Schmidt in 1887.[2] Both Schmidt and later Borrmann in 1907 described this condition as lethal.[9] The incidence rate of this tumor has remained relatively constant, accounting for 0.04% of all breast tumors.[4] and approximately 8% of mammary sarcomas.[5] Several terms have been used to describe this malignant condition such as hemangioendothelioma,[3] haemangioblastoma,[6] hemangiosarcoma[7,8] and metastasizing angioma.[9–11]

The usual clinical presentation of angiosarcoma is a painless smooth mass, but in approximately 17% of cases the tumor may appear with red discoloration and apparent bruising of the overlying skin.[12] PAB carries very poor prognosis with a five-year survival rate of 8–50%.[13] The mean latency ranges from 5 to 6 years. Distant metastases have been observed in the lungs, skin, liver, bones, CNS, spleen, ovary, lymph nodes and heart.[14,15] The angiosarcoma may relapse in the same breast especially after conservative surgery and postoperative radiation.[16]

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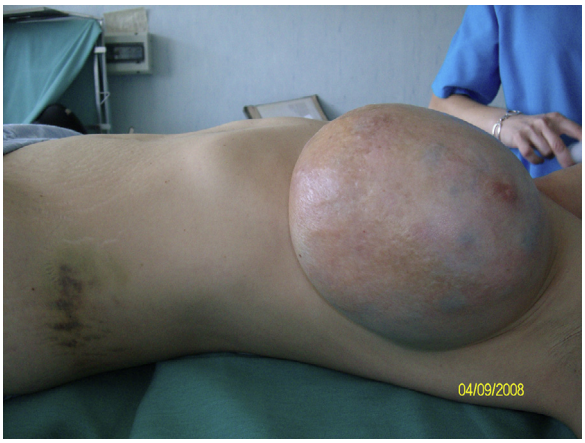

Fig. 1 – A bluish red discoloration of the overlying skin of the breast and in the middle of the abdominal region.

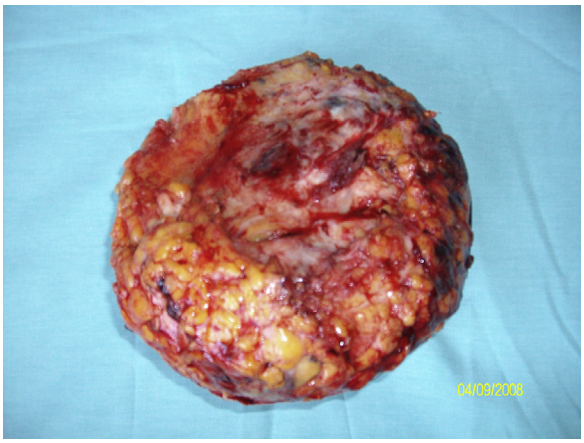

Fig. 2 – Completely surgically excised tumor.

We report an uncommon clinical case of an advanced breast angiosarcoma, without distant metastases, but associated with disseminated intravascular coagulation (DIC) by consumption coagulopathy, known as the Kasbach–Merritt syndrome.

2. Case report

The patient was a 42-year-old woman who had a tumor in the left breast for three years. During this period the patient refused any treatment for her disease and no such treatment was done. In the last two months, before the current hospitalization, the tumor rapidly grew and the overlying skin colored in red. The patient was admitted to the National Cancer Center Sofia on August 2, 2007, in a critical condition. The physical examination revealed a hard, mobile, painless mass located in the central region of the left breast. On palpation the tumor dimensions were 18/16/14 cm and had sharp limits. No enlargement of axillary lymph nodes, no nipple retraction and no skin thickening were found. The right breast was normal. The overlying skin of the affected one had bluish red discoloration (Fig. 1). Similar red discoloration was seen on other parts of the body such as the back, the legs and the middle of the abdominal region (Fig. 1). X-ray mammography of the left breast showed a big tumor mass of 18.5 cm in diameter, without spiculae or microcalcifications. Ultrasound showed a liquid area in the central zone of the tumor. The tumor marker CA15-3 was within the referenced limits. Distant metastases were not established. On admission, the coagulation tests showed exhausted haemostatic potential with low platelet count, no detectable fibrinogen and no coagulation of APTT and PT. There were high levels of D-dimmers. These laboratory findings, together with the clinically manifested bleeding diathesis with big suffusions on the skin in the sites of venepunctures, were interpreted to be caused by disseminated intravascular coagulation (Table 1). D-dimers were measured using commercial antibody-based assay.

Substitution therapy was performed with 7 units of platelet concentrate, 10 ml/kg fresh frozen plasma and 2 units of red blood cell concentrate. The response to the therapy was highly satisfactory. Platelet counts reached 70×10^9/l, fibrinogen increased to 3.5 g/l and APPT and PT were normalized. The surgery was performed with no excessive bleeding. The normal range of platelets was reached on the next day after surgery with no additional substitution therapy (Table 1). The tumor was completely excised en bloc with the first level axillary lymph nodes due to their being visibly enlarged (Fig. 2).

The mastectomy specimen showed bruising of the skin and a well-defined tumor. The cut surface of the tumor was soft, spongy, bluish, hemangioma-like, with necroses and hemorrhages as results of numerous dilated blood vessels (Fig. 3).

Microscopic examination of the tumor revealed a mixed structure – it contained areas of endothelial tufting with papillary formation and solid spindle cell components, corresponding to poorly differentiated angiosarcoma – Grade 3 (Fig. 4). Hemorrhages, necroses, cellular pleomorphisms and mitotic figures were also observed. By means of immunocytochemistry, endothelial marker CD 31 was used, and it strongly stained the sarcoma cells (Fig. 5).

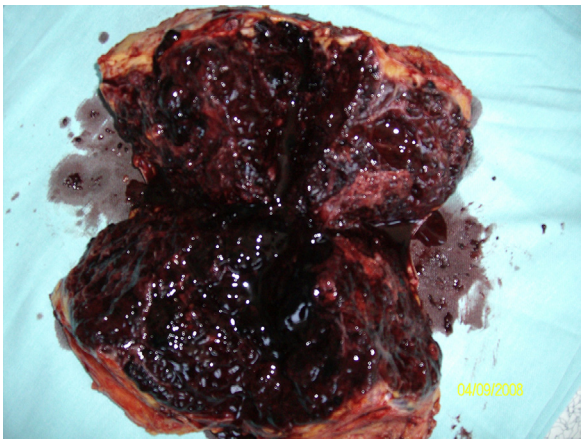

Fig. 3 – Necrosis and hemorrhages in the central tumor area.