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Ein ungewöhnlicher Fall mit einer möglichen Erklärung für fatale Thrombozytopenie.

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ABSTRACT

In the last fifteen days, a 22 year old male has been experiencing breathlessness on exertion, ecchymosis, jaundice, and signs of worsening right heart failure. Auf der körperlichen Untersuchung wurde festgestellt, dass er einen mittleren diastolischen Murmel in der Trisepsregion und einen austretenden systolischen Murmel in der Lungenregion hatte. Die Untersuchung der Knochenmarkhistopathologie zeigte eine erhöhte Anzahl von Megakaryocyten. Berichte über routinemäßige Untersuchungen waren üblich. Eine einzelne große intrakardiale Masse, die aus dem rechten Atrium stammt, wurde auf einer Computertomographie (CT)-Scannung entdeckt. Dies führte zu einer dynamischen Blockade des In- und Ausflusses des rechten Herzens. Die verbundene fatale thrombocytopenie zeigte keine Reaktion auf intravenöse Steroide oder platelettransfusionen. Die Patientin konnte nicht operiert werden, da sie einen sehr niedrigen Plateletcount hatte und während ihres Krankenhausaufenthalts starb, bevor eine Excisionbiopsie durchgeführt werden konnte.

Es wurde keine pathologische Autopsy durchgeführt. This is a rare case because the fatal thrombocytopenia that was seen here was caused by mechanical effects like frictional and shear force, which can be explained by the physical presence of a large intracardiac mass that blocks flow.

Key words: Frictional force, intra cardiac tumor, mechanical force, shearing force, thrombocytopenia

INTRODUCTION

Es ist offensichtlich, dass kardiovaskuläre Tumoren mit schwerer thrombozytopenie und Blutgerinnsel äußerst selten sind.[1-6] Thrombocytopenia in Verbindung mit einem intrakardialen Tumor hat verschiedene Ursachen. Die bloße Anwesenheit einer großen Masse im Herzen wirkt sich stark auf die Blutzellen aus und kann schwere Verletzungen a den Plateletten verursachen. Es kann manchmal zu tödlicher thrombozytopenie führen. According to a few studies, an improvement in platelet count after cardiac mass removal is supported.[4–6]

CASE REPORT

Over the course of 15 days, a 22 year old male experienced progressive breathlessness on exertion, increasing jaundice, easy bruising, and petechial rash. Es gab keine Vorgeschichte von Epistaxis, Gingivalblutungen, Hämaturie, Hemoptysis, Fieber, Gewichtsabnahme, neurologischen Anzeichen oder einem Thromboembolismus. Es gab keine familiäre Geschichte von Blutgerinnseln oder jüngere Exposure a heparin oder heparinoids. In der Untersuchung hatte er eine Blutdruck von 110/70 mmHg, eine Herzfrequenz von 90/min und eine regelmäßige Herzfrequenz. He war betrunken. He had ecchymosis and his jugular venous pressure was elevated 9 cm above Louis' angle. As a sign of right ventricular inflow obstruction, he experienced facial puffiness and soft tender hepatomegaly. Durch eine Herzauskultation haben wir einen langen, mittleren diastolischen Murmur im Trikotraum und einen starken phasenmäßige Ausscheidung von systolischem Murmur der Klasse III/VI ohne Ausscheidungsklick im Pulmonalbereich. In der peripheral smear zeigte er eine schwere thrombozytopenie, während seine direkten Bilirubinwerte, die Gesamtzahl der Leukocyten und Neutrophilen erhöht waren.

Bone marrow study showed normal cellularity and erythroid prominence (myeloid:erythroid ratio 1:1, normoblastic erythropoesis, normal orderly myelopoesis, no blast prominence, increase in megakaryocytes number with few hypoblastic forms and prominence of histiocytes). Chest radiograph and electrocardiography were normal. Abdominal ultrasound showed uniform congestive hepatomegaly with normal size spleen. Color Doppler ruled out deep vein thrombus in abdomen or lower limbs. Trans-thoracic echocardiography [Figure 1] showed dilated right atrium, and hypertrophied and dilated right ventricle. There was a solitary, giant pedunculated, lobulated, homogenous mobile mass obstructing right ventricular inlet and outlet, exactly similar to a large right atrial myxoma originating

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from fossa ovalis.

Contrast enhanced computerized tomography (CT) of chest and abdomen showed a single large non enhancing soft tissue density occupying the right atrium, right ventricle and right ventricular outflow tract, measuring 10 × 7.5 mm, and extending into inferior venacava with dilated of right atrium and right ventricle [Figure 2]. Mediastinum and lung parenchyma were normal. There was a mild bilateral pleural effusion, homogenous hepatomegaly and mild ascites, with other viscera as normal. Hence, the possible radiological diagnosis was of an intra cardiac thrombus. Anti phospholipids antibody, anti systemic lupus erythematosus (SLE) antibodies and anti platelet factor 4 (PF4) antibodies were negative. He was treated with empirical intravenous antibiotics, intravenous methyl prednisolone and 20 units of single donor platelet transfusion. Neither anticoagulants nor thrombolytic agents were used during the treatment. Excisional biopsy was not possible because of continuous decline in platelet counts, and none of the serial follow

up platelet count levels were above 15, 000/mm^3. On the 15th day of hospitalization, he died of cardiac arrest just one day after an episode of melena. We could not manage pathological autopsy because of the lack of consent.

DISCUSSION

The possible causes of thrombocytopenia related to cardiac pathology are as follows:

Space occupying lesions

1. 1. Tumoren des Herzens wie myxoma[7,8], angiosarcoma, rhabdomyoma, lymphoma, carcinoid tumor, papilläres fibroelastoma, renales Zellkarzinom und metastabile germzelltumoren. Use of drugs, autoimmune response, disseminated intravascular coagulation, thrombotic thrombocytopenic purpura, hemorrhage associated with extensive transfusion, paraneoplastic syndrome, and idiopathic thrombocytopenic purpura are the postulated mechanisms of thrombocytopenia associated with cardiac tumors. Es besteht die Möglichkeit, dass eine große Masse oder ein Tumor im Herzen vorhanden ist, die die Blutzellen mechanisch durch Reibung und Reibung schädigt.[2,2,5] 2. Thrombose und Pflanzen
2.
3. .

Here, thrombocytopenia may be due to mechanical damage, autoimmunity and infection.

Stenotic lesions

1. Native valvular stenotic lesions.
2. Mechanical valves.
3. Devices like coils and occluder.

In these conditions, the possible cause of thrombocytopenia

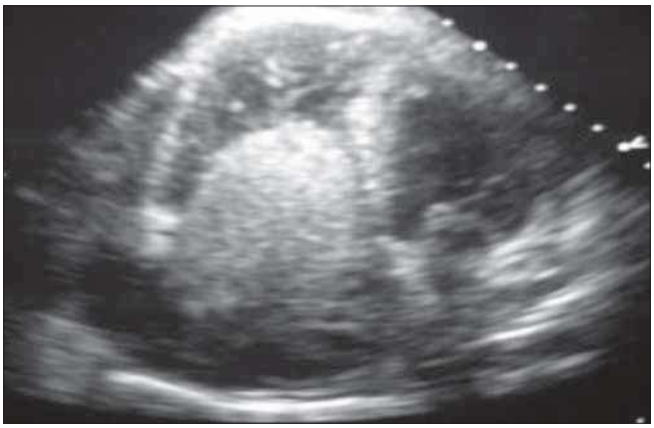

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Figure 1: 2 Dimensional Echocardiograph in apical 4 chamber view (A4C) shows a right atrial, single, mobile, homogenous, large 7 × 10 cm mass, without satellite lesions, and causing dynamic obstruction to the right ventricular inflow and outflow tract

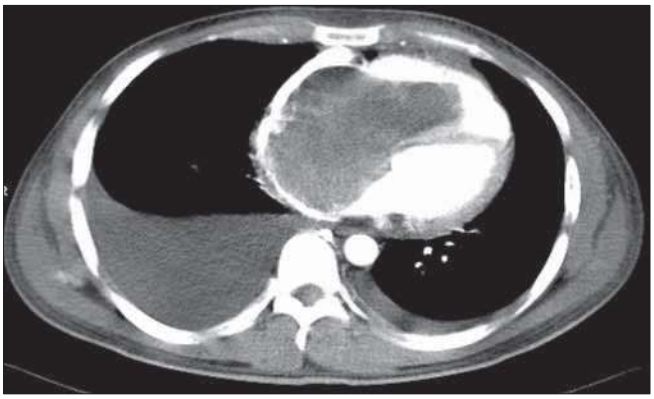

Figure 2: Computerized tomography with contrast shows a single and large contrast non enhancing intra cardiac mass occupying right atrium and right ventricle with right ventricular outflow tract and inferior venacava extension. This typical description is consistent with intra cardiac thrombus

and anemia is mechanical friction. Amongst the cardiac tumors, myxoma is most common followed by angiosarcoma and lymphoma. Both these tumors are more common in males, and have special predilection for the right side of the heart, particularly the right atrium. Right heart failure is the most common presenting symptom in these tumors. Further, angiosarcoma is much more common whereas primary lymphoma of the heart is rare. Angiosarcoma is the second most common primary cardiac tumor after myxoma, and also the commonest tumor of the right atrium. Rhabdomyoma is more frequently seen in children, and it originates from the myocardium of either ventricle. Rhabdomyosarcoma is common in children and adolescents, but rare in adults.[1,2] Papillary fibroelastoma is the most common of the tumors involving the heart valves, and is usually seen in the elderly and not associated with thrombocytopenia. Renal cell carcinoma was excluded because of normal abdominal ultrasound and computerized tomography (CT). Carcinoid tumor mainly presents as a tricuspid valve dysfunction. Right heart failure due to intra cardiac blood flow obstruction is a relatively frequent complication of large cardiac tumors.[7-9]

Thrombocytopenia is associated with both, benign[1-3] and malignant[4-6] cardiac tumors. Thrombocytopenia has also been found to be associated with other hematologic disorders such as anemia,[8,9] and erythrocytosis;[5] however, in our case thrombocytopenia was an isolated finding. The exact etiopathogenesis of thrombocytopenia associated with solid cardiac tumors is unclear. Idiopathic thrombocytopenic purpura (ITP) is the most common cause of thrombocytopenia in females in the age group of 15 to 50 years of age, and is defined as thrombocytopenia in apparently healthy patients without other associated causes. The defining features of ITP include thrombocytopenia, microangiopathic hemolytic anemia, neurological signs, renal abnormalities and fever. Thus, both idiopathic thrombocytopenic purpura (ITP) and thrombotic thrombocytopenic purpura (TTP) are ruled out in our case as there are no evidence of end organ damage, micro angiopathic hemolytic anemia and no response to steroid therapy. Coomb´s test was negative. A negative anti systemic lupus erythmatosus (SLE) antibodies report excluded Evan syndrome. Normal lactate dehydrogenase and normal haptoglobin levels exclude hemolysis. The sudden cardiac death in the index case can be explained by sudden obstruction to inflow or outflow tract, pulmonary thromboembolism or due to massive intracranial bleed. Thrombocytopenia due to impaired production is ruled

out as bone marrow shows increased megakaryocytes. The role of antibody induced thrombocytopenia was excluded as there was no response to steroids given for two weeks.

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CONCLUSION

Es ist äußerst ungewöhnlich, einen intrakardialen Tumor mit thrombozytopenie zu haben. Es wurden mehrere mögliche etiopathologische Faktoren genannt, aber die exakte Ursache ist noch nicht bekannt. Despite steroids and platelet transfusions, some cases fail to respond. In such situations exists a significantly low platelet count, so that the most important investigations like biopsy or definitive surgeries cannot be done. Mechanical forces like frictional and shearing stress exerted by the mere presence of intra cardiac tumors are rare but occasionally cause significant damage to the blood cells, which may be the cause of this fatal thrombocytopenia. To establish a causal association, more detailed studies are needed.

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