Scientific letters

Castleman Disease Infiltrating Great Vessels and Right Atrium

Enfermedad de Castleman que infiltra grandes vasos y aurícula derecha

To the Editor,

Castleman disease, or angiofollicular lymph node hyperplasia, is a rare lymphoproliferative disorder of unknown etiology and pathogenesis. Its importance lies in the fact that it has been associated with human immunodeficiency virus (HIV) and human herpes virus 8, as well as other neoplastic diseases such as Kaposi's sarcoma, lymphomas, and POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes).

We present the case of a 66-year-old man with no known cardiovascular risk factors who complained of dyspnea upon moderate exertion, together with a weight loss of 4 to 5 kg over a 6-month period. In the physical examination, a grade 2/6 systolic murmur was detected at the pulmonary site (left second intercostal space) on auscultation. A chest X-ray showed no evidence of cardiomegaly or signs of pulmonary congestion. Transthoracic echocardiography revealed the presence of mild pulmonary stenosis with a peak pressure gradient of 23 mmHg and, on the parasternal short-axis view, there was a marked concentric thickening of the ascending aorta, which exerted pressure on the pulmonary artery. In view of the echocardiographic findings, we performed a magnetic resonance study (Figs. 1 and 2), which revealed a soft tissue mass surrounding the aortic root and ascending thoracic aorta and occupying the lumen of the pulmonary trunk. The mass extended toward the right pulmonary artery, occupying its entire lumen and infiltrating the right atrium as well. Following the administration of an intravenous contrast medium, there was a marked enhancement of the mass that revealed the extensive vascularization of the lesion. Positron emission tomography/computed tomography was performed to rule out the existence of lesions at other sites. The patient was referred to cardiac surgery for resection of the mass. Following median sternotomy, the pericardium was opened and an infiltrative process involving the aortic root and ascending aorta, pulmonary trunk, and right atrium was identified. Because of the close proximity of the lesion to the great vessels, only partial resection was possible, concluding with layered closure using a Peri-Guard repair patch. The final pathology report concluded that the lesion consisted of a polyclonal lymphoid proliferation that was compatible with the plasma cell variant of Castleman disease. As the mass had only been partially resected, the decision was made to administer neoadjuvant therapy with the anti-CD20 monoclonal antibody, rituximab. The patient has responded favorably to treatment and 1 year later is asymptomatic.

Follow-up computed tomography scans show a considerable reduction of the size of the mass. Any association of this case with HIV or with other neoplastic diseases was ruled out.

Castleman disease has two variants with highly different prognoses and treatments: multicentric and unicentric, the latter constituting 70% of the reported cases. The most common sites are chest (70%), neck, abdomen, retroperitoneum, and pelvis. Histological criteria define two types: the hyaline vascular variant (the more prevalent type) and the plasma cell variant (which accounts for only 10% to 20% of the cases).

Figure 1. Axial “black-blood” magnetic resonance image showing a soft tissue mass surrounding ascending thoracic aorta, extending to and occupying the lumens of pulmonary trunk and right pulmonary artery (arrows).

Figure 2. Coronal “black-blood” magnetic resonance image showing a soft tissue mass occupying the entire right pulmonary artery lumen (arrows).
Unicentric Castleman disease is a generally benign disorder that is usually classified as belonging to the hyaline vascular type. It is found in young adults, and most of the patients are asymptomatic. In contrast, the multicentric form is found in older adults and is usually associated with the plasma cell variant, a systemic disease with generalized lymphadenopathy, hepatosplenomegaly, fever, and night sweats; in addition, it is frequently associated with HIV infection.2

The treatment of choice of the unicentric form is surgical, and complete resection is curative in most cases. If the lesion cannot be resected completely, the prognosis with partial resection is also favorable, and the patient may remain asymptomatic for years. Other therapeutic options include preoperative embolization, radiotherapy, and chemotherapy. The experience with the use of rituximab in unicentric Castleman disease is limited, although there are reports of cases in which promising results were obtained in patients with unresectable disease or in whom partial resection was performed. In the multicentric form, surgical resection is not sufficient, and the association of radiotherapy and chemotherapy is necessary.3

Castleman disease is a rare disorder, the diagnosis of which requires a high degree of suspicion, due to the absence of specific clinical or radiological findings; the definitive diagnosis is based on the pathological study. It should be considered in the differential diagnosis of any mediastinal mass.4

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REFERENCES
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Safety and Efficacy of Endothelial Progenitor Cell Capture Stent in ST-Elevation Acute Myocardial Infarction. GENIA Study

Seguridad y eficacia del stent capturador de células progenitoras de endotelio en el infarto agudo de miocardio con elevación del ST. Estudio GENIA

To the Editor,

The Genous® stent (OrbusNeich, Fort Lauderdale, Florida, United States) is made of stainless steel coated with murine anti-CD34 monoclonal antibodies, an antigen present on the surface of endothelial progenitor cells. Because of this characteristic, circulating cells of this type are attracted to the stent and attach to the struts, resulting in prompt formation of a layer of functional endothelium in less than 2 weeks.1 In a highly prothrombotic clinical situation such as ST segment elevation acute myocardial infarction (STEAMI), fast endothelialization of the stent could hypothetically reduce the risk of thrombosis and the need for new target vessel revascularization (TVR).

The aim of our study is to evaluate the safety and efficacy of the Genous® stent in patients with STEAMI undergoing primary angioplasty.

This is a prospective observational study carried out between June 2008 and July 2010, including 139 consecutive patients undergoing primary angioplasty with implantation of one or more Genous® stents. Patients who were hospitalized in cardiogenic shock, those in recovery from cardiac arrest, and patients with a formal contraindication for dual antiplatelet therapy for at least 1 month were excluded. The regimen for antithrombotic and antiagulant therapy followed the recommendations of European guidelines for the management of acute myocardial infarction. Patients were followed up by telephone contact.

Cardiac death was defined as death due to a cardiac cause, an unknown cause, or a procedure-related cause. Clinical restenosis was established on the presence of anginal symptoms associated with >50% stenosis in the segment covered by the stent and the adjacent 5 mm.

The patients’ baseline characteristics and the procedure-related characteristics are summarized in Tables 1 and 2.

All patients received dual antiplatelet therapy, which lasted for 1 year in 88 patients (65.2%). Mean follow-up was 538 (334.72) days. Five patients died during follow-up (3.6%); one death was due to noncardiac cause related to gastric neoplasm at 2 years following the procedure (cardiac mortality 2.9%, noncardiac mortality 0.7%), 2 patients died suddenly at home at 1 year and 2 years of follow-up, and 2 patients died during hospitalization (1 due to cardiac rupture and 1 due to cardiogenic shock and multiorgan failure).

Table 1
Baseline Characteristics of Patients With ST Segment Elevation Acute Myocardial Infarction Treated With a Genous® Stent

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Patients (n=139)</th>
<th>(%)</th>
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<tbody>
<tr>
<td>Age, years</td>
<td>64±13.5</td>
<td></td>
</tr>
<tr>
<td>Males</td>
<td>104 (74.8)</td>
<td></td>
</tr>
<tr>
<td>Smokers</td>
<td>67 (58.2)</td>
<td></td>
</tr>
<tr>
<td>Hypertension</td>
<td>69 (49.6)</td>
<td></td>
</tr>
<tr>
<td>Hyperlipidemia</td>
<td>66 (47.5)</td>
<td></td>
</tr>
<tr>
<td>Diabetes mellitus</td>
<td>24 (17.3)</td>
<td></td>
</tr>
<tr>
<td>Previous AMI</td>
<td>13 (9.4)</td>
<td></td>
</tr>
<tr>
<td>Chronic renal failure</td>
<td>10 (7.3)</td>
<td></td>
</tr>
<tr>
<td>Previous revascularization surgery</td>
<td>1 (0.7)</td>
<td></td>
</tr>
<tr>
<td>Previous PCA</td>
<td>14 (10.1)</td>
<td></td>
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<tr>
<td>Maximum TpI ng/mL</td>
<td>93.1±87</td>
<td></td>
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<tr>
<td>Multivessel disease</td>
<td>28 (20.1)</td>
<td></td>
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<tr>
<td>Ejection fraction</td>
<td>52.3±10.8</td>
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</tr>
</tbody>
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AMI, acute myocardial infarction; PCA, percutaneous coronary angioplasty; TpI, troponin I.

Data are expressed as mean±standard deviation or no. (%).