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Title: Rosai-Dorfman disease: Presentation of a clinical case

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Body: Introduction: Rosai-Dorfman disease (RDD) is a rare histiocytic proliferative process, infrequent and idiopathic, benign with painless cervical or systemic linfandenopathy, fever, leukocytosis, polyclonal hypergammaglobulinemia and accelerated erythrocyte sedimentation. In a low percentage, there are extranodal manifestations. The histological diagnosis consists of filling of sinusoids in ganglia with linfocytes, plasmatic cells and histiocytes positive for S100, CD68, absence of CD1a and emperipolesis. Case report: 49-year-old female patient, who was a heavy smoker is presented with severe dyspnoea, three months evolution and chest oppression sensation, with edema of face, neck and arms. The erythro sedimentation was accelerated (90 mm/hr). Chest x-rays showed mediastinum widening and tumor in the pulmonary right hilum. The computerized tomography showed right hilum hyperdensity of 7 cm. The bronchoscopy with biopsy showed right bronchi infiltrate with tumoral aspect. The hysthopathological findings revealed hysthiocyte proliferation and others with eosinophilic cythoplasma with fibrosis and lymphoid cells. Emperipolesis and plasmatic cells where identified, with Russel bodies. With the diagnosis 120 mg a day of 'attack dose' was initiated, continuing with 40 mg/day during 6 months with favorable response reducing the size of the lesion. Discussion: In this case, the manifestation of the disease was dyspnoea produced by the bronqueal obstruction and compression of the superior vena cava due to the hilum infiltrative histiocytic mass, so our initial probable diagnosis was a bronchogenic cancer. This extranodal manifestation is infrequent, but it is to be taken into account as part of the pulmonary and mediastinal pathology.