The growth rate of paragangliomas is slow and they are usually benign. The criteria of malignancy is not defined by the histopathology of the tumour, but whether they spread to adjacent organs, due to metastasis or recurrence, which are found in 5-10% of the total.[4] The treatment of choice is surgical resection, which is considered risky due to its anatomical location: the close relationship with the vascular-nervous structures and hypervascularity of the tumour. The main postoperative complication is sensory or motor deficit caused by nerve injury to adjacent structures. Radiation therapy is indicated for inoperable cases or as a complement to surgery after partial resection. However, this therapy does not usually completely eradicate the tumour.[5]

References

1. Gehman KE, Currie I, Ahmad D, Parrent A, Rizkalla K. Desmoid tumour of the thoracic outlet: an unusual cause of thoracic outlet syndrome. Can J Surg. 1998;4:404-6.
2. Sergeant G, Gheysens O, Seynaeve P, Van Cauwelaert J, Ceuppens H. Neurovascular compression by a subpectoral lipoma. A case report of a rare cause of thoracic outlet syndrome. Acta Chir Belg. 2003103:528-31.
3. Yeow KM, Hsieh HC. Thoracic outlet syndrome caused by first rib hemangioma. J Vasc Surg. 200133:1118-21.
4. Mondragón-Sánchez A, Montoya Rojo G, Shuchleib-Chaba S. Tumor de cuerpo carotídeo (paraganglioma). An Med Asoc Med Hosp ABC. 200348:233-6.
5. Maier W, Marangos N, Laszig R. Paraganglioma as a systemic syndrome: pitfalls and strategies. J Laryngol Otol. 1999113:978-82.

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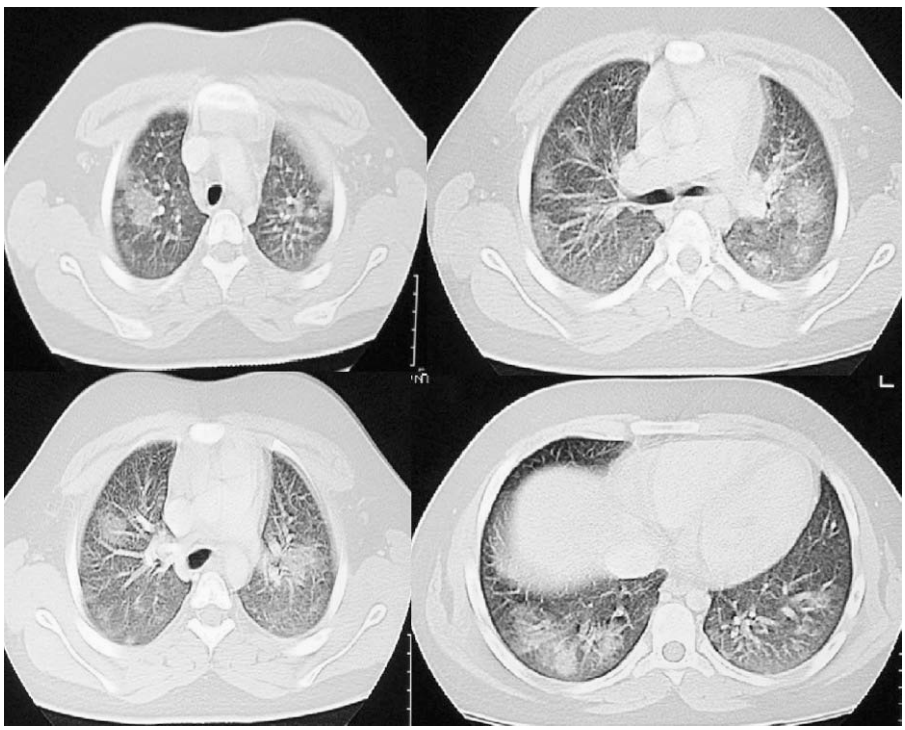
Idiopathic Pulmonary Haemosiderosis in Childhood: A Good Response to Systemic Steroids, Inhaled Hydroxychloroquine and Budesonide

Hemosiderosis pulmonar idiopática en la infancia: buena respuesta al tratamiento con esteroides sistémicos, hidroxicloroquina y budesonida inhalada

To the Editor:

Idiopathic pulmonary haemosiderosis (IPH) is a rare and potentially lethal cause of diffuse alveolar haemorrhage. It is characterised by the presence of changing lung infiltrates, haemoptisis and ferropenic anaemia; with no systemic or renal associated symptoms.[1,2] Its clinical presentation varies from fulminant haemoptisis and acute respiratory failure to insidious clinical respiratory symptoms or refractory ferropenic anaemia.[1,3,5]

We present the case of a 10 year old boy, with no previous respiratory symptoms, with irritative cough and progressive dyspnoea of one year of evolution accompanied by haemoptisis during the last 6 months. On exploration the most outstanding symptoms were obesity (BMI 27) and paleness of skin and mucous membranes. Additional tests showed ferropenic anaemia (haemoglobin 10.3 mg/dl, haematocrit 32%). Functional renal, liver, ions, coagulation and urine tests were all normal. Sweat test results 12 meq/l. Sputum culture was negative and Mantoux test 0 mm. On chest X-ray it was possible to see thickening of the hilum with bilateral interstitial infiltration predominantly of the lower lobes. The CAT scan can be seen in annex 1. Heart studies were normal. Negative results were obtained for immunoglobulin, complement,



RF, ANA, ANCA, antiDNA, antiMBG, IgA ATT and precipitins to cow's milk. Studies of pulmonary function showed a restrictive pattern: forced spirometry: FEV1 79%, FVC 80%, FEV1/FVC 90%, negative bronchodilation and plethysmography: TLC 67%, FVC 67%, RV 21%, with moderate decrease of CO diffusion capacity. No macroscopic alterations were seen on fibrobronchoscopy; on bronchoalveolar lavage a bloody serous fluid was obtained with abundant red blood cells (49%) and haemosiderophages (41%). Cultures for moulds, bacteria and mycobacterium were negative.

The patient was diagnosed with IPH in the absence of any findings to suggest other respiratory conditions which could be causing the alveolar bleeding. Treatment was initiated with oral prednisone (2 mg/kg/day), inhaled budesonide (400 μg/day), hydroxychloroquine (7 mg/kg/day), ferrous sulphate and inhaled salbutamol as needed. After 2 months oral prednisone is decreased, and the patient presented two episodes of slight haemoptisis; however, he reports better resistance to physical stress and the anaemia disappears. At 14 months, the patient has no symptoms and is receiving 10 mg of prednisone every 48 hours, hydroxychloroquine is decreased and is eventually suspended 2 years after it was begun; 10 months after prednisone is discontinued. At present, 3 years after diagnosis, the patient is on inhaled budesonide (400 μg/day) and has not had any new exacerbations, only presenting dyspnoea on exertion. All analytical, radiological and lung function tests are normal and the patient has not suffered any adverse effects due to medication.

It is difficult to give recommendations for IPH treatment. The series of cases published have a limited number of patients and their response to drugs is variable. Systemic steroids as first line of treatment in acute cases seem to control bleeding, improve X-ray images and decrease morbimortality.[1,3-5] Long term benefits are less evident; although their prolonged use is related to greater survival, a decrease in exacerbations and less lung fibrosis.[1-4,6] The adverse effects due to prolonged use of systemic corticosteroids in infancy and the risk of recurrences when dosage is decreased make it necessary to find alternative treatments.[3,5,6]

The effectiveness of hydroxichloroquine has been documented, both during the acute phase as during maintenance after corticosteroid failure.[2-5] Furthermore, there is no evidence to indicate what is the appropriate dose, when to begin and when to stop treatment. Cyclophosphamide has also been successfully used during the acute phase and during maintenance, especially in threatening situations that have not responded to corticosteroid treatment.[2] There are also publications reporting the benefits of azathioprin to control symptoms in the long term and so reduce the dose of corticosteroids.[2,3,5] High doses of inhaled corticosteroids have also been used to decrease the dosage of systemic corticosteroids, although neither is there sufficient evidence to support their general use.[1,2,5,6]

To conclude, we wish to point out that a prolonged treatment with a combination of inhaled and systemic corticosteroids and hydroxichloroquine achieved an appropriate response in the case we have presented. The appearance of new episodes of haemoptisis when prednisone was first decreased prolonged the duration of treatment. The good response seen in our patient, although this was an isolated case, is an indication that it is possible to consider using this combined therapy for IPH.

Funding

None.

Conflict of Interest

The authors affirm that they have no conflict of interest.

Appendix 1

Chest CAT: Multiple bilateral diffuse infiltrates, some of pseudonodular morphology, with ground glass areas and small sized hilar adenopathies.

References

1. Ioachimescu OC, Sieber S, Kotch A. Idiopathic pulmonary haemosiderosis revisited. Eur Respir J. 2004;24:162-70.
2. Nuesslein TG, Teig N, Rieger CHL. Pulmonary haemosiderosis in infants and children. Paediatr Respir Rev. 2006;7:45-8.
3. Saeed MM, Woo MS, MacLaughlin EF. Prognosis in pediatric idiopathic pulmonary hemosiderosis. Chest. 1999;116:721-5.
4. Le Clainche L, Le Bourgeois M, Fauroux B, Forenza N, Dommergues JP, Desbois JC, et al. Long-term outcome of idiopathic pulmonary hemosiderosis in children. Medicine. 2000;79:318-26.
5. Kabra SK, Bhargava S, Lodha R, Satyavani A, Walia M. Idiopathic pulmonary hemosiderosis: clinical profile and follow up of 26 children. Indian Pediatr. 2007;44:333-8.
6. Kiper N, Gocmen A, Ozcelik U, Dilber E, Anadol D. Long-term clinical course of patients with idiopathic pulmonary hemosiderosis (1979-1994): prolonged survival with low-dose corticosteroid therapy. Pediatr Pulmonol. 1999;27:180-4.

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Secondary Amyloidosis with Renal Involvement in an Adult Patient with Cystic Fibrosis

Amiloidosis secundaria con afectación renal en paciente adulto con fibrosis quística

To the Editor:

Cystic fibrosis (CF) is a genetic disease from which survival has been increasing steadily over recent decades.[1] Other complications associated with CF have also been increasing, such as secondary amyloidosis, which is associated with chronic inflammatory processes. Amyloidosis is a systemic disease characterised by the extracellular deposition of fibrillar proteins. Secondary AA amyloidosis consists of fibrils of protein A, an acute phase reactant produced by hepatocytes. Renal involvement is common in this condition. Secondary AA amyloidosis is a recognised complication of CF (mainly in patients with a long evolution of the disease and poor disease control), but very rare. Its incidence is not known in CF and it is associated with poor prognosis. In most cases it presents with proteinuria, thryomegaly, and/or hepatosplenomegaly.[2] Furthermore, amyloidosis with renal involvement is frequent and evolves into kidney failure in a relatively short time (months or years), which is associated with a poor prognosis.[3]