Angioedema as a single manifestation of carcinoid syndrome in a bronchial carcinoid tumor


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ABSTRACT

Background: The association of bronchial carcinoid tumours with carcinoid syndrome is extremely rare especially in the absence of metastatic disease, and the angioedema is not a typical sign of this syndrome.

Methods and results: We report the case of a 39-year-old woman referred to our allergy department with recurrent episodes of angioedema. The aetiological study of angioedema did not show evidence of hypersensitivity to common inhalants, food allergens and latex. C1-inhibitor, C3, C4, C1q, proteinogram and immunoglobulins (IgA, IgG, IgM) all were normal. TSH determination gave normal results, too. Faecal analyses for parasites were negative. The haemogram showed moderate leucocytosis and hypocromic microcytic anaemia. The thoracic radiography showed a mediastinal node image in the right paratracheal region. Histology analyses of the samples were diagnostic of a typical carcinoid tumor. Levels of 5-hydroxyindolacetic acid (5-HIIA) were slightly increased. A superior lobectomy was performed and no new episodes of angioedema appeared after surgical intervention.

Conclusions: We report the first case of typical bronchial carcinoid tumour, without metastatic disease, with angioedema as a single manifestation of carcinoid syndrome.

In our knowledge, only one case of Quincke’s edema as part of typical carcinoid syndrome has been reported, in a case of primary midgut carcinoid tumor with metastatic disease to liver. It is very important to include complementary tests, as thoracic radiography, in the routine study of angioedema to reject malignant diseases.

Key words: Angioedema. Carcinoid syndrome. Carcinoid tumor. Thorax radiography.

RESUMEN

La presencia de síndrome carcinoide asociado a tumores bronquiales es poco frecuente, sobre todo en ausencia de enfermedad metastásica; y el angioedema no es una manifestación típica de dicho síndrome.

Métodos y resultados: Presentamos el caso de una paciente de 39 años de edad con episodios recurrentes de angioedema.

En el estudio etiológico de angioedema no se evidenció hipersensibilidad frente a inhalantes, alimentos ni látex. La determinación de fracciones séricas de complemento (C3, C4, C1q y C1-inhibidor) e inmunoglobulinas mostró resultados normales. Los va-
lores de TSH estaban, asimismo, dentro de la norma- lidad. En análisis de parásitos en heces fue negativo. En el hemograma se apreciaba una leucocitosis mo- derada y una anemia microcítica e hipocromía. La radiografía de tórax mostraba una imagen no- dular mediastínica a nivel paratraqueal derecho. El estudio histológico fue diagnóstico para carciñoide típico. Los niveles de ácido 5-hidroxindolacético (5-HIIA) en orina de 24 horas, estaban discretamen- te elevados. A la paciente se le practicó una lobecto- mia superior derecha, no volviéndose a presentar nuevos episodios de angioedema tras la interven- ción.

Conclusiones: Presentamos el primer caso de tu- mor carciñoide bronquial típico, sin enfermedad me- tastásica asociada, con angioedema como única ma- nifestación de síndrome carciñoide.

Sólo tenemos conocimiento de un caso de ede- ma angioneurótico de Quincke asociado al síndrome carciñoide en un caso de tumor primario intestinal con metástasis hepáticas. Creemos que es impor- tante incluir determinados exámenes complementa- rios, como la radiografía de tórax, en el estudio de rutina del angioedema para descartar enfermedades malignas subyacentes.

associated to this clinical situation. It is of interest the measurement of metabolite 5-HIIA in a 24 hour urine collection (Noris). This situation is possible in extra-intestinal locations such as lung, gonads and the retroperitoneum. Carcinoid tumors arising in the lung specifically produce serotonin, gastrin, adrenocorticotropic hormone and histamine, probably involved in the genesis of angioedema in this case. Therefore, bronchial carcinoid tumors with carcinoid syndrome are extremely rare, and it has been reported that gastrointestinal carcinoids usually produce the syndrome only when they spread to the liver, because from there they can release tumour products into the systemic circulation avoiding liver breakdown, and metastasize to the lungs, bones and other organs.

But, as said, the development of the syndrome is possible in the absence of metastasis, if the tumour discharges the active products directly into the systemic circulation and circumvents hepatic metabolism. This situation is possible in extraintestinal locations such as lung, gonads and the retroperitoneum.

Carcinoid tumors that are characterised by a low rate of serotonin production are usually characterised by a low rate of serotonin production. In our patient’s case, the levels of 5-HIIA (5-hydroxyindoleacetic) were slightly elevated, although normal for serotonin. However, there are atypical forms of carcinoid syndrome in which those biochemical markers are normal and it is necessary to determine other metabolites of tryptophan as 5-HTP (5-hydroxytryptophan) or 5-HT (serotonin).

In conclusion, we report the first case of typical bronchial carcinoid tumor, without metastatic disease, with angioedema as a single manifestation of carcinoid syndrome. So, it is very important to include complementary tests in the routine study of angioedema to reject malignant diseases. In our case, the thoracic radiography was the basis of the diagnosis of recurrent angioedema due to bronchial carcinoid tumor.

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REFERENCES