EDITORIAL COMMENT

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Thymoma paraneoplastic syndrome awareness

Thymomas are rare malignant epithelial tumors of the thymus. Usually presenting as indolent nodules of the thymic fat pad, they have the potential to grow into surrounding structures and dedifferentiate into more aggressive forms with metastatic potential.

As the thymus is related to the immune response, its malignancies can be associated with a myriad of paraneoplastic auto-immune syndromes (PNS), being miastenia gravis the most frequently found in association with thymomas.[1]

In this issue of PJCTVS, Gomes et al bring us a case series of Good's syndrome. This and other PNS are often underdiagnosed, still when investigating the cause for recurrent infections, hypogammaglobulinemia has been found in up to 11% of thymoma patients either prior to the diagnosis or after curative intent surgery.

Thymoma associated autoimmune syndromes can affect several organs and systems but the most frequent can generally be categorized into 3 groups: neuromuscular, hematological and dermatological. However, a recent systematic review of 507 surgically treated thymoma patients with PNS found 123 different types of paraneoplastic thymoma associated syndromes of which nephrotic syndrome represented 5.7% of cases.[1]

Upfront surgical management of resectable thymic tumors is recommended only if a complete resection with clear margins is feasible.[2] Successful complete resection of thymoma is defined as a complete thymectomy from phrenic nerve to phrenic nerve and from the diaphragm to the upper poles of the thymus, including any involved structures.[3]

In Zhao's review, there was heterogeneity of surgical treatment, not amenable to detailed analysis. In spite of that, surgical resection is associated to complete or partial symptom resolution in 76% of cases. Non-miastenic PNS seamed to confer a worse prognosis to thymoma patients, and PNS recurrence occurred in 34% of cases within the first 2 years of follow-up and was associated to a worse survival.[1]

Professionals treating thymic malignancies must do so in a multidisciplinary team with experience in this particularly rare tumor management, including dedicated thoracic surgeons. For its high prevalence, myasthenia Gravis should be excluded in thymoma patients[2]. Additionally, symptom awareness to other PNS should prompt early investigation and customized treatment, both before and after thymectomy, as PNS can be associated to thymoma recurrence, conferring a worse prognosis.

REFERENCES

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