

AperTO - Archivio Istituzionale Open Access dell'Università di Torino

Severe episodic dyspnoea and abnormal flow-volume loop. Extramedullary plasmacytoma (EMP).

This is the author's manuscript

Original Citation:

Availability:

This version is available <http://hdl.handle.net/2318/127639> since

Published version:

DOI:10.1136/thx.2008.100024

Terms of use:

Open Access

Anyone can freely access the full text of works made available as "Open Access". Works made available under a Creative Commons license can be used according to the terms and conditions of said license. Use of all other works requires consent of the right holder (author or publisher) if not exempted from copyright protection by the applicable law.

(Article begins on next page)



ANSWER

Thorax 2009;64;264
doi:10.1136/thx.2008.100024a

Updated information and services can be found at:
<http://thorax.bmj.com/cgi/content/full/64/3/264>

These include:

References

This article cites 5 articles, 1 of which can be accessed free at:
<http://thorax.bmj.com/cgi/content/full/64/3/264#BIBL>

Email alerting service

Receive free email alerts when new articles cite this article - sign up in the box at the top right corner of the article

Notes

To order reprints of this article go to:
<http://journals.bmj.com/cgi/reprintform>

To subscribe to *Thorax* go to:
<http://journals.bmj.com/subscriptions/>

20. **Stolz D**, Christ-Crain M, Morgenthaler NG, *et al*. Copeptin, C-reactive protein, and procalcitonin as prognostic biomarkers in acute exacerbation of COPD. *Chest* 2007;**131**:1058–67.
21. **Parr DG**, White AJ, Bayley DL, *et al*. Inflammation in sputum relates to progression of disease in subjects with COPD: a prospective descriptive study. *Respir Res* 2006;**7**:136.
22. **Pinto-Plata V**, Toso J, Lee K, *et al*. Profiling serum biomarkers in patients with COPD: associations with clinical parameters. *Thorax* 2007;**62**:595–601.
23. **Dahl M**, Vestbo J, Lange P, *et al*. C-reactive protein as a predictor of prognosis in chronic obstructive pulmonary disease. *Am J Respir Crit Care Med* 2007;**175**:250–5.
24. **Fogarty AW**, Jones S, Britton JR, *et al*. Systemic inflammation and decline in lung function in a general population: a prospective study. *Thorax* 2007;**62**:515–20.
25. **Man SF**, Connett JE, Anthonisen NR, *et al*. C-reactive protein and mortality in mild to moderate chronic obstructive pulmonary disease. *Thorax* 2006;**61**:849–53.
26. **Shaaban R**, Kony S, Driss F, *et al*. Change in C-reactive protein levels and FEV₁ decline: a longitudinal population-based study. *Respir Med* 2006;**100**:2112–20.
27. **Fabbri LM**, Romagnoli M, Crobetta L, *et al*. Differences in airway inflammation in patients with fixed airflow obstruction due to asthma or chronic obstructive pulmonary disease. *Am J Respir Crit Care Med* 2003;**167**:418–24.
28. **Zietkowski Z**, Kucharewicz I, Bodzenta-Lukaszyk A. The influence of inhaled corticosteroids on exhaled nitric oxide in stable chronic obstructive pulmonary disease. *Respir Med* 2005;**99**:816–24.
29. **Rose G**. Environmental health: problems and prospects. *J R Coll Physicians Lond* 1991;**25**:48–52.
30. **Wallis EJ**, Ramsay LE, Haq I, *et al*. Coronary and cardiovascular risk estimation for primary prevention: validation of a new Sheffield table in the 1995 Scottish health survey population. *BMJ* 2000;**320**:671–6.
31. **Jackson R**. Updated New Zealand cardiovascular disease risk-benefit prediction guide. *BMJ* 2000;**320**:709–10.
32. **Jackson R**. Guidelines on preventing cardiovascular disease in clinical practice. *BMJ* 2000;**320**:659–61.
33. **Anderson KM**, Odell PM, Wilson PW, *et al*. Cardiovascular disease risk profiles. *Am Heart J* 1991;**121**(1 Pt 2):293–8.
34. **de Torres JP**, Campo A, Casanova C, *et al*. Gender and chronic obstructive pulmonary disease in high-risk smokers. *Respiration* 2006;**73**:306–10.
35. **Fletcher C**, Peto R. The natural history of chronic airflow obstruction. *BMJ* 1977;**1**:1645–8.

Pulmonary puzzle

ANSWER

From the question on page 210

This case describes an extremely rare condition of extramedullary plasmacytoma (EMP) hidden in the middle mediastinum, giving no systemic signs but causing severe central airway narrowing detectable by the pattern of the flow-volume loop. The constant expiratory and inspiratory flow limitation is consistent with severe fixed narrowing of the central airway.¹ Accurate analysis of the flow-volume curve pattern at the onset of symptoms would have given rise to a suspicion of tracheal compression several months earlier.

Fixed intrathoracic airway obstruction is usually due to intramural infiltration (post-endotracheal intubation, recurrent polychondritis, primitive tracheobronchial neoplasia and amyloidosis) or to extrinsic compression (intrathoracic goitre, thymoma, lymphoma).² Mediastinal localisation of plasma-cytoma is very uncommon, usually presenting as a large mass visible on the chest radiograph.³ In this case, EMP was hidden in the middle mediastinum and could only be detected on the chest CT scan.

Transbronchial biopsy was consistent with IgG lambda plasmacytoma. Multiple Russell bodies on PAS staining were indicative of cytoplasmic inclusions of immunoglobulins. The results of immunohistochemical staining are shown in fig 1.

EMP is a plasma cell neoplasm of soft tissue without bone marrow involvement or other systemic characteristics of multiple myeloma, representing about 3% of all plasma cell neoplasms.⁴ It can be differentiated from reactive plasmacytoma and plasma cell granuloma or lymphoma (MALT, marginal and immunoblastic) by the expression of specific cell surface markers.⁵

This case suggests that the pattern of the flow-volume loop may give a hint of central airways narrowing caused by hidden masses not visible by traditional procedures.

Thorax 2009;**64**:264. doi:10.1136/thx.2008.100024a

REFERENCES

1. **Kryger M**, Bode F, Antic R, *et al*. Diagnosis of obstruction of the upper and central airways. *Am J Med* 1976;**61**:85–93.
2. **Ernst A**, Feller-Kopman D, Becker HD, *et al*. Central airway obstruction. *Am J Respir Crit Care Med* 2004;**169**:1278–97.
3. **Miyazaki T**, Kohno S, Sakamoto A. A rare case of extramedullary plasmacytoma in the mediastinum. *Intern Med* 1992;**31**:1363–5.
4. **Alexiou C**, Kau RJ, Dietzfelbinger H, *et al*. Extramedullary plasmacytoma. *Cancer* 1999;**85**:2305–14.
5. **Guidelines Working Group of the UK Myeloma Forum (UKMF)**. Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. *Br J Haematol* 2004;**124**:717–26.

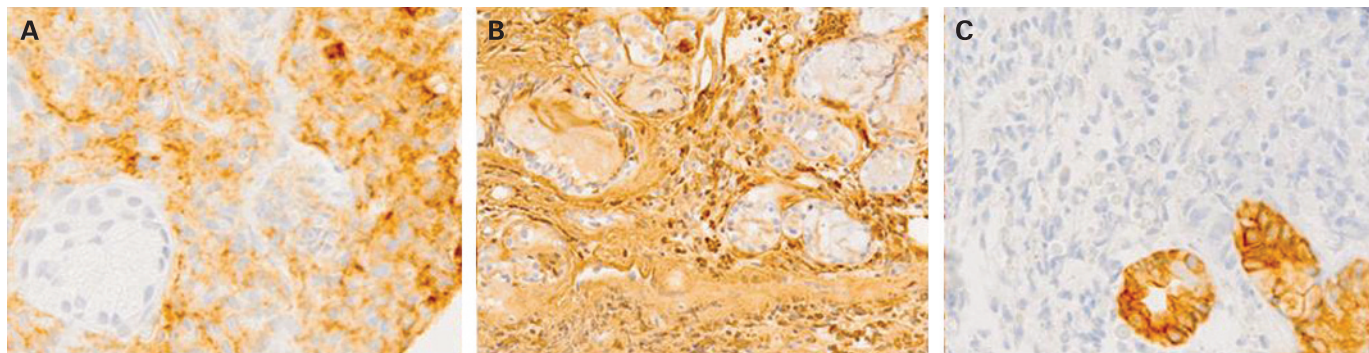


Figure 1 Immunohistochemical analysis of the proliferating cells phenotypically characterised to be (A) CD138+, CD79a+, (B) clgG lambda+ (weak), (C) clgG kappa- and cytokeratin AE1/3-. Staining for clgM, clgD, clgA, clgE, CD3 and CD45RO were all negative. Since there were very few non-neoplastic (clg-kappa+) plasma cells and small lymphocytes, these findings were suggestive of IgG lambda plasmacytoma.