Bullous pemphigoid: combined diagnostic criteria are still needed

Dear Editor

There is no doubt that the pathogenetic pathway of bullous pemphigoid (BP) is undiscovered in its whole spectrum. Literature is being enriched with further experience on combined diagnostic procedures. In our study we used combined methods to establish diagnosis of BP in 40 patients (75.1 ± 11.8 years), 17 men and 23 women. 15 healthy volunteers and 15 patients with pemphigus vulgaris were enrolled as controls. Enrollment of patients was based on the presence of typical clinical and histological picture, as well as positive direct or indirect immunofluorescence (DIF or IIF). Blood sera were examined by BP180NC16A and BP230 ELISAs, prior to initiation of treatment. Anti-BP180ELISA had a sensitivity of 67.5%, while sensitivity of anti-BP230 ELISA was 40%. Total sensitivity (for both antigens) was 77.5%, a value that was competitive to DIF. Specificity of anti-BP180 and anti-BP230 assay was 100%, while diagnostic accuracy was 81.4% and 65.7%, respectively. When both ELISAs were used in combination, specificity was 100% and diagnostic accuracy was 87.14%. Levels of autoantibodies were higher in patients with widespread lesions. Atypical features do not allow clinical picture of BP to become a definite criterion. The percentage of atypical BP presentations is estimated to be around 40% \( Joly \) et al, has considered significant the co-existence of four clinical criteria: age > 70 years, absence of residual scars, absence of mucosal lesions and lesions on the head and neck \( Joly \). In that study, 75% of patients had positive at least 3 out of 4 criteria \( Joly \). In our study, 77.5% of patients were positive for all clinical criteria, while 22.5% of patients had 3 out of 4.

Histological findings are diagnostic in 50-70% of cases \( 1,3 \). Histology is connected to the age of lesion. Old blisters or lesions with poor inflammatory infiltrates may be misdiagnosed.

DIF is often performed in inadequate specimens, resulting in a pathology report with the conclusion “suggestive”. IIF depends on the phase of BP. Chaidemenos et al, comparing clinical, histological and immunofluorescence findings, observed a variety of results without intracorrelation. The most important conclusion was that 25% of patients would have been under a wrong diagnosis if IIF was not performed \( 1 \). Immunoblotting is a method with high potential in identifying pathogenic antigens. However, it isn’t a practical method for everyday use. On the other hand, ELISA is an easy technique, able to detect specific circulating autoantibodies and quantitate their levels. Sensitivity increases with ELISAs that detect several BP180 extra or intracellular epitopes or proteins of hemidesmosomes \( 4 \).

In conclusion, combined diagnostic criteria are still needed for the diagnosis of BP.

References

Dear Editor,

The presence of a lateral neck mass often creates a diagnostic challenge. Despite the aid of imaging methods, the clinical doctor and surgeon should be aware of even unusual entities that must be included in the differential diagnosis.

A 72-year-old Greek female was admitted to a tertiary referral center, for further evaluation and management of an asymptomatic lateral neck mass, that was first noted 6 months before, during routine clinical examination. Patient’s history included a right and a left thyroid lobectomy twenty and ten years ago, respectively, due to multinodular goiter. Physical examination revealed a deeply situated, difficult to palpate, soft, non-pulsatile, non-tender mass located in the right anterolateral area of the neck (level III). Routine laboratory exams were unremarkable with normal thyroid gland function. Ultrasonography was followed by MRI that also revealed an atractoid mass in the right parapharyngeal space, medial to the common carotid artery and internal jugular vein, extending from the level of the oropharynx to the level of the larynx. No enlarged lymph nodes were detected. A clear diagnosis was not evident and therefore surgical exploration of the neck was decided. The mass was approached through a lateral neck incision. Dissection and complete removal was performed after conservation of the major neck blood vessels and cranial nerves. Postoperative period was uneventful. Permanent histology was consistent with normal thyroid tissue.

Ectopic thyroid tissue is defined as any thyroid tissue found in a location other than the normal, pretracheal region, with a prevalence of approximately one in 100,000 to 300,000 persons. Ectopic thyroid gland generally occurs in the midline of the neck as a result of abnormal median migration, and its presence lateral to the midline is rare. Ninety percent of ectopic thyroid tissue cases are found in the area of the foramen cecum as lingual thyroid, with other known anatomic areas including intratraheal, submandibular, oropharyngeal, lingual thyroid, with other known anatomic areas including intratraheal, submandibular, oropharyngeal, lingual thyroid, but often located elsewhere. The differential diagnosis of a lateral neck mass includes metastatic lymph node, submandibular gland tumor, branchial cleft cyst, carotid body tumor, lipoma, neurofibroma, sebaceous cyst, parathyroid adenoma, primary soft tissue tumor, infection (including tuberculosis, syphilis), cystic hygroma and ectopic thyroid tissue. The presence of a lateral neck mass is a diagnostic challenge for the clinical doctor. Useful diagnostic tools include US, MRI, CT, Angiography, Scintiscan and FNAB. Surgical excision is the treatment of choice, with selective or modified radical neck dissection in the rare case of thyroid carcinoma with metastatic lymph nodes.

In conclusion, ectopic thyroid tissue in the lateral neck, despite its low prevalence, should be included in the differential diagnosis of any lateral neck mass. Surgeons should be aware of the existence of an ectopic thyroid gland in unusual locations, especially whenever imaging results are incompatible with other pathological entities.

References

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Keywords: ectopic thyroid tissue, lateral neck mass, diagnosis, surgery

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