

Differential diagnosis of adrenal insufficiency in a patient with APS syndrome and Rosai-Dorfman disease (RDD)

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Introduction:

Primary adrenal insufficiency could be a rare manifestation of antiphospholipid syndrome (APS) and may be a consequence of bilateral adrenal hemorrhage. Rosai-Dorfman disease (RDD) is sinus histiocytosis with massive lymphadenopathy, a rare histiocytic proliferative disorder with a distinctive microscopic appearance. The retro-peritoneum is an infrequent site of involvement as well as extranodal sites.

Case presentation:

- a 39 year old female patient with diagnosed APS 3 years ago treated with anticoagulant therapy and glucocorticoids presented with fatigue, loss of appetite and abdominal cramps.
- One year prior admission an abdominal ultrasound showed acalculose cholecystitis along with lymphadenopathy and a right adrenal gland enlargement 3 cm in diameter, while left was described as normal.
- At the time of admission primary adrenal insufficiency was diagnosed while other hormone tests were normal and prednisone was replaced with hydrocortisone.
- In the mean time abdominal CT revealed again acalculose cholecystitis but now both adrenal glands were enlarged and hypodensely structured, left 2,6x3,9, right 3,8x1,8 cm with numerous enlarged lymph nodes in the area of hepatoduodenal ligament largest being 3,3 cm in diameter.
- Surgical removal of gallbladder was performed along with extirpation of lymph nodes and patohistological finding showed sinus histiocytosis.
- Postoperative synacthen test indicated low basal cortisol production with no response to stimulation and hydrocortisone therapy was continued.

Conclusions:

Differential diagnosis included adrenal hemorrhage associated with APS or Rosai-Dorfman disease (RDD) which can be associated with APS (only two cases reported presently) but retroperitoneal location is extremely rare let alone extranodal sites. However, since CT imaging characteristics showed no signs of hemorrhage, thrombosis or embolism of adrenal glands, it is entirely possible that bilateral enlargement and adrenal insufficiency was a result of sinus histiocytosis infiltration as a sign of RDD.

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