A rare case of bilateral adrenocortical carcinoma with metastasis in a young male

Dr. Dixit Varma and Dr. Nirupam Konwar Baishya

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Abstract

Although adrenal tumours are very common, majority of them are benign lesions. Adrenocortical carcinoma is a rare endocrine malignancy and carries a poor prognosis. They are often unilateral and detected in older age groups. This report describes a case of bilateral adrenocortical carcinoma in a 30-year-old male who presented with cervical lymphadenopathy that demonstrated malignant cells on fine needle aspiration (FNA) cytology. Ultrasound and CECT examinations of the abdomen detected bilateral adenal tumours. Imaging characteristics were correlated with CT guided FNA of the lesion and diagnosed as adrenocortical carcinoma. This case highlights the importance of considering malignant adrenal aetiology in bilateral adrenal masses as such cases are rare in literature.

Keywords: Adrenocortical carcinoma, Fine needle aspiration (FNA), poor prognosis

Introduction

With non-specific hyperplasia, metastasis from the head and neck region and lymphoma being the common causes of cervical lymphadenopathy, the possibility of adrenal carcinoma as the cause of cervical lymphadenopathy is often overlooked. Adrenal masses are often incidentally detected on routine radiological investigations. Bilateral adrenal tumours are infrequent and some of the common causes reported include pheochromocytoma, metastatic tumours, lymphoma, nonfunctioning cortical adenoma primary aldosteronism and Cushing’s syndrome [1]. Adrenocortical carcinomas are rare and the estimated incidence is thought to be around one to two per 1,000,000 per year [2]. Adrenocortical carcinomas are bilateral in 10% of cases [3]. The rarity of the condition and contradiction of biopsy in early investigation, as suggested by recent guidelines pose a delay and difficulty in the diagnosis of adrenocortical carcinomas.

Case

The patient is a 30-year-old male who presented to a tertiary care hospital with complaints of progressive neck swelling for 3 months. There was no dysphagia, pain, fever or weight loss. There was also no history of heat or cold intolerance or tachycardia. The patient has no other significant past medical history. There is no family history of malignancy. His Blood pressure was 120/70 mmHg and his pulse was 78/min. There were no signs or symptoms of adrenal hormonal hypersecretion. Physical examination revealed multiple nodular swellings over bilateral regions of the neck. Lab parameters were unremarkable except for leukocytosis 28.0 × 109/L with neutrophilia. Free plasma metanephrines were within normal limits. Ultrasonography of the neck revealed multiple well defined enlarged heteroechoic lymph nodes with hypoechoic areas within, (suggestive of necrosis) and increased colour uptake on colour doppler study in bilateral regions of the neck at cervical levels II to IV (Figure 1). Fine needle aspiration (FNA) cytology was done from the enlarged lymph nodes and was suggestive of metastatic adenocarcinoma. An ultrasound of the abdomen was performed on the patient, which showed two large well defined heteroechoic mass lesions in bilateral suprarenal regions, measuring up to 111 mm x 105 mm x 115 mm on the right side and 112 mm x 104 mm x 111 mm on the left side, with a mild increase of colour uptake on colour doppler study (Figure 2). The rest of the ultrasound findings were unremarkable. The patient was then advised a CECT of the abdomen and a CT guided FNA from the adrenal glands. CECT revealed large heterogeneously enhancing mass lesions in bilateral suprarenal regions (Figure 3a). The right lesion is abutting the posteroinferior surface of the right lobe of the
liver with effacement of fat planes between them. There is an ill-defined hypoenhancing area in the body of the pancreas (Figure 3b) and sclerotic areas in L4, S1 vertebral bodies and ala of sacrum, all of which are suggestive of neoplastic infiltration. A heterogeneously enhancing enlarged perigastric mesenteric lymph node is also noted. The CT findings were thus, in favour of malignant neoplastic aetiology. HRCT of the thorax was done to rule out primary lung neoplasms and lung involvement. The findings were suggestive of interstitial pattern of lung disease. FNA from the adrenals was done under CT guidance and revealed malignant cells suggestive of adrenocortical carcinoma. However, following the FNA, the patient started feeling giddiness and sweating along with a drop in blood pressure. The patient was admitted to ICU and started on inotropes, however, the blood pressure could not be maintained and he eventually met with cardiorespiratory arrest and could not be revived.

Fig 1: Ultrasonography of cervical lymph region demonstrating enlarged Level III lymph node on the right side, showing a hypo-echoic within (arrow), suggestive of necrosis

Fig 2: Ultrasonography images showing large well defined heteroechoic mass lesions (white arrow heads) arising from supra renal regions bilaterally. The right kidney (red arrow) and left kidney (black arrow) are noted inferior to the lesions.

Fig 3: The coronal reformatted portal venous phase (a) and axial portal venous phase computed tomography images demonstrating bilateral adrenocortical carcinomas. There is heterogenous enhancement of the mass lesions. Hypoenhancing areas (red arrow) are noted in the pancreatic parenchyma suggestive of infiltration.
Discussion
Adrenocortical carcinomas are rare and aggressive tumours involving the adrenal glands. In About 40-60 per cent of cases, the patient presents with signs and symptoms of adrenal hormone excess. As adrenocortical carcinomas are often large, about one-third of the patients present with symptoms of pressure effect due to the tumour, like abdominal fullness and flank pain. About 20-30 per cent of cases are incidentally detected. Adrenocortical carcinomas are more usually sporadic and unilateral. Hereditary cases are associated the Beckwith-Wiedemann syndrome, Li-Fraumeni syndrome, Carney complex, MEN-1 syndrome and McCune-Albright syndrome. Detection of bilateral adrenal masses warrants a metastatic workup to rule out other primary malignancies. Common primary sites include the lung, stomach, oesophagus and liver. Adrenocortical carcinomas are found in older patients with a median age of 61 years. Women are more commonly affected than men, with a ratio of about 2:1. Overall, Adrenocortical carcinomas have a poor prognosis and the most important prognostic factor is the stage at the time of diagnosis. Hence early detection and consideration of the possibility of malignant aetiology is important in management.

Initial evaluation of adrenocortical carcinomas includes thorough physical examination and physical examination for signs and symptoms of hormonal excess. Initial hormonal assessment is crucial. Staging should include at least CT or MRI of the abdomen and CT of the chest. Evaluation for metastasis should be guided by clinical suspicion.

The ideal treatment for non-metastatic adrenocortical carcinoma is complete resection of the tumour. In cases where complete resection is not possible, debulking surgery may be considered to decrease the hormonal secretion. In patients with widespread distant metastatic deposits, the primary tumour can be treated with external beam radiation as a palliative measure. The increased incidence of tumour recurrence even after complete resection of the tumour has pointed out the importance of adjuvant therapy. Mitotane is the treatment option for inoperable tumours. It is the only drug approved by the U.S. Food and Drug Administration and European Medicine Executive Agency for the treatment of adrenocortical carcinoma. Mitotane is administered alone or in conjunction with chemotherapy agents.

Conclusion
Adrenocortical carcinomas are rare neoplasms with a poor prognosis. The most important prognostic factor is the stage at the time of presentation and hence early detection is important. In cases with cervical lymphadenopathy of metastatic aetiology, it is important to keep adrenocortical carcinomas as a differential. Although rare, it is also important to keep adrenocortical carcinomas as a differential while evaluating bilateral adrenal masses. FNA should also be done with caution as it can lead to hemodynamic instability.

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Dr. Dixit Varma was the primary author. Dr. Nirupam Konwar Baishya also contributed equally to this work.

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