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Short Reports

Cushing's Syndrome in Infancy Due to Pituitary Adenoma

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Abstract. An 11-month-old male with Cushings syndrome due to a pituitary adenoma is presented. Additional findings of bilateral lung and kidney cystic disease – chance association or new syndrome?

Key words: Cushing's syndrome – Infants and children – Pituitary adenoma – Cystic lung disease – Cystic renal dysplasia

Etiologies of Cushing's syndrome vary according to age. Adrenal hyperplasia predominates in children over 7 years of age [1]. Under age 7 years, adrenal tumors predominate [2]. The most common causes under age 1 year are adrenal neoplasms, diffuse nodular adrenal hyperplasia, and ectopic ACTH syndrome [3]. The rarest etiology during infancy is pituitary adenoma with only one reported case [4].

We report a second case with emphasis on the radiographic findings. In addition, bilateral cystic lung and renal disease accompanied hypercortisolism.

Case Report

This 11-month-old boy was referred for evaluation of probable Cushing's syndrome. Physical examination revealed typical Cushingoid facies, truncal obesity, acne vulgaris, and facial hair. His weight and head circumference exceeded the 95th percentile with length at the 50th percentile. Striae and hyperpigmentation were absent; eye and genitalia exams were normal. A palpable right lower quadrant mass extended from the umbilicus to the anterior superior iliac crest.

Remarkable past history included bilateral lung cysts (Fig. 1) resected at age 5 weeks due to worsening respiratory status. Pathological diagnosis was benign congenital lung cysts. Subsequent evaluation of an enlarging left lower quadrant abdominal mass with ultrasound revealed a multiloculated cystic kidney (Fig. 2 A) which was subsequently resected at age 6 months (Fig. 2 B). Pathological diagnosis was Potter Type II A multicystic kidney.

Laboratory data at admission included: normal serum electrolytes, markedly elevated excretion of 17-hydroxy corticosteroids and increased serum cortisols. Plasma ACTH was upper limits of normal. Positive radiologic studies included retarded bone age (6 standard deviations below normal), abdominal sonography and computerized cranial tomography (CCT). Ultrasound examination of the right lower quadrant mass revealed cystic nephromegaly identical in appearance to the resected left multiloculated cystic kidney (Fig. 2 A, B) without recognizable adrenal gland abnormalities; computerized body tomography (CBT) (Fig. 3 A) was confirmatory. Skull roentgenograms revealed a questionable sella turcica erosion. CCT showed a suprasellar mass (Fig. 3 B) consistent with a pituitary adenoma. The pituitary lesion was resected and the pathological diagnosis was mixed chromophobe and basophilic adenoma with subsequent immunohistochemical confirmation. The patient remains stable without signs of hypercortisolism at the present.

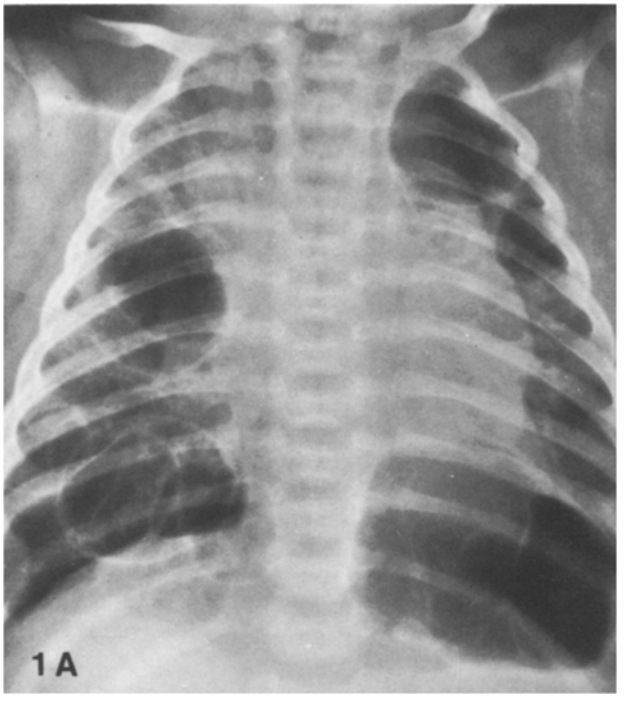

Fig. 1. Anteroposterior chest roentgenogram shows bilateral multiple varying sized cystic lesions

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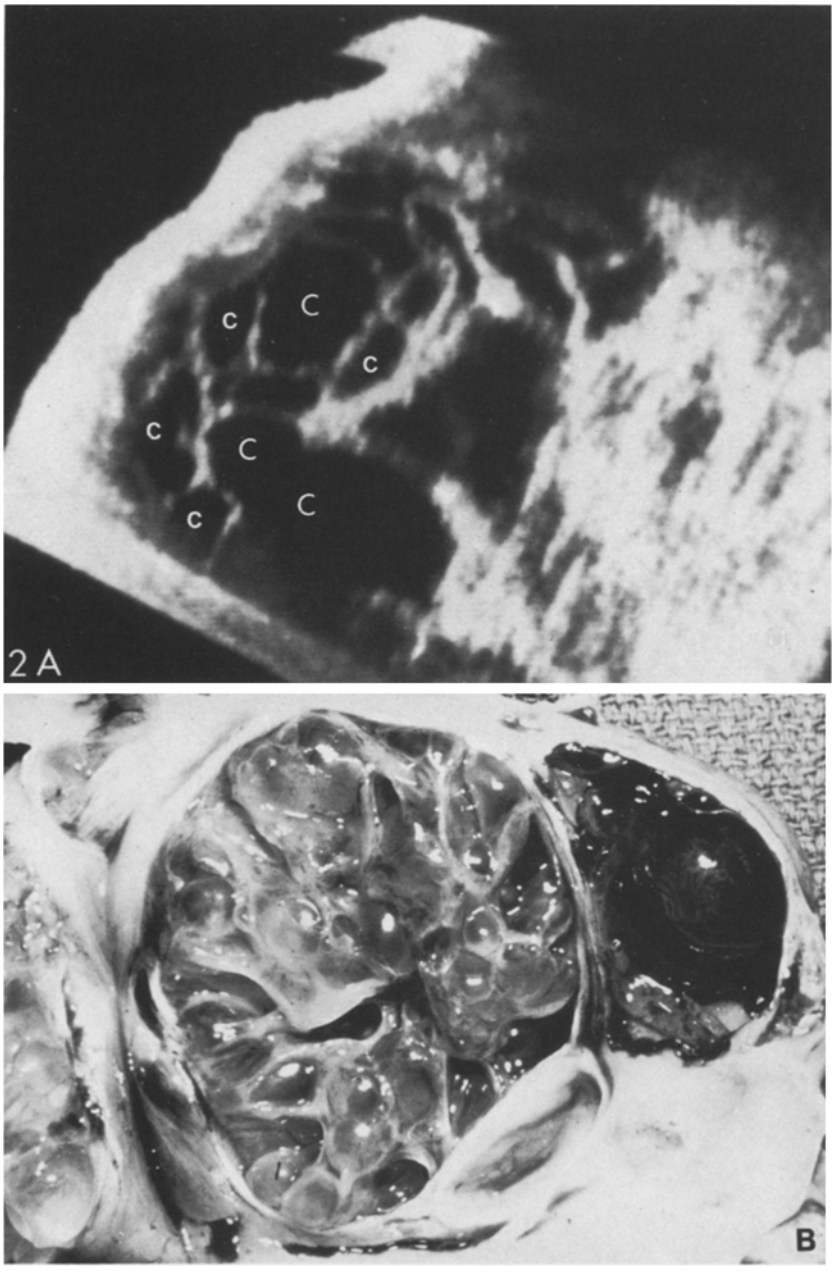
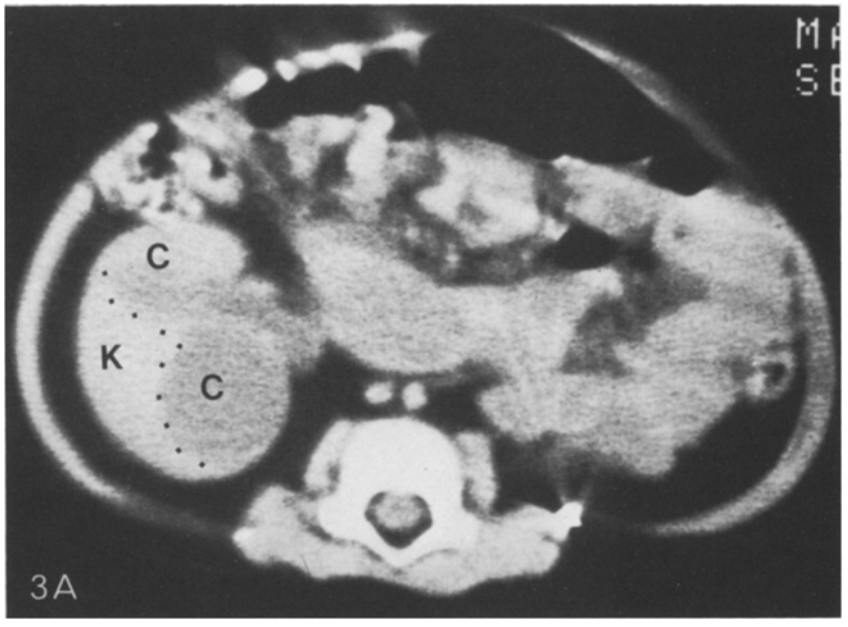


Fig. 2. A Transverse supine real-time ultrasound scan reveals huge multiloculted left cystic kidney (C-cyst). **B** Surgical specimen showing cystic replacement of left kidney

Discussion

60 cases of Cushing's syndrome in infancy have been reviewed [3–5]. 75% of reported cases were female. Adrenocortical tumors constitute the majority (48/60) with approximately equal incidence of carcinoma and adenoma. Nine patients had adrenal hyperplasia and two patients had ectopic ACTH syndrome. The only previously reported infant with a pituitary adenoma was diagnosed at age 8 months and died in the immediate postoperative period [4]. Cushing's syndrome in infancy has a high mortality rate with only 62% postoperative survival as compared with 85% survival of older children and adults [6].

In this infant as in others, Cushing's syndrome was not suspected until the clinical manifestations were apparent. If laboratory data indicates an adrenal etiology for hypercortisolism, ultrasound and/or CBT is indicated. Abdominal ultrasound was initially chosen for evaluation of an abdominal mass in this patient with subsequent additional sonographic exclusion of adrenal mass or enlargement; CBT revealed similar findings. If an intracranial lesion is suspected, skull roentgenograms and CCT are war-



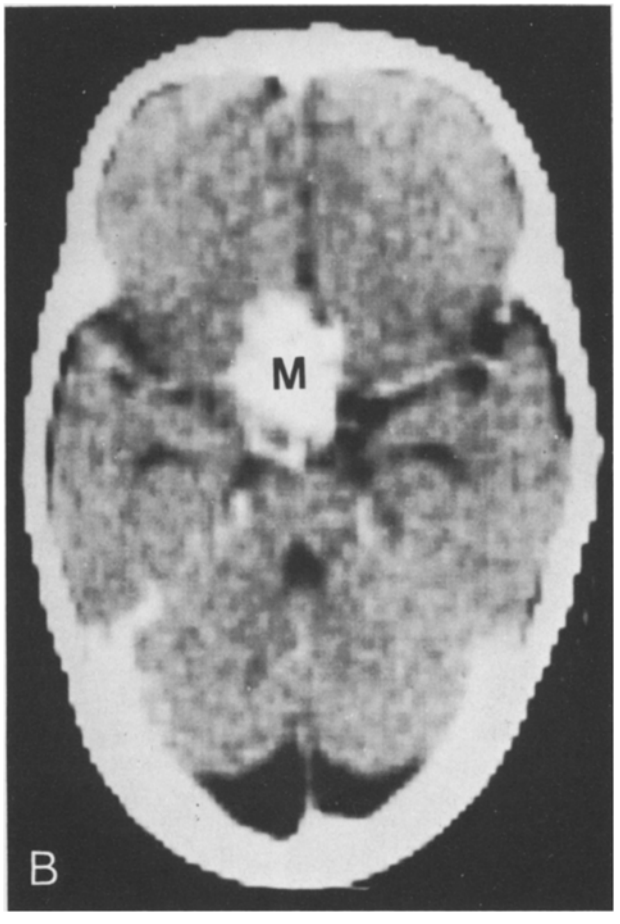


Fig. 3. A CBT scan reveals huge multiloculated right cystic kidney (C-cyst; K-kidney). No adrenal abnormality seen. **B** CCT scan demonstrates enhancing suprasellar mass (M-mass)

ranted. Skull radiographs, including sellar tomograms may be negative and CCT may be normal with pituitary microadenomas [7]. Cerebral angiography and/or pneumoencephalography may next be indicated if a suspected hypophyseal lesion remains undetected.

An association between Cushing's syndrome and cystic disease of lung and kidneys remains unknown. We have not learned of any similar reported combinations so the triad of lung cysts, renal cysts and pituitary adenoma may represent either chance occurrence or a new entity.

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