

## Case Summary Section

This section is a summary of the clinical characteristics, imaging features, pathology findings, and treatment and prognosis for the diagnosis with representative radiology and pathology images from your patient. A case with an excellent case summary may be selected for an online publication with AIRP.

Radiologists and radiation oncologists should not claim as their intellectual property that which is not theirs. Plagiarism or the use of others' work without attribution is unethical. Please review the [American College of Radiology - Code of Ethics](#).

### **Representative Images – This is the most critical component to a successful case summary.**

Please include all appropriate imaging modalities in your submission. Imaging planes should match gross images to best appreciate radiologic-pathologic correlation.

### **Examples of clean representative images with and without annotation (JPEG format only):**

**ALL images must be anonymized, cropped, and completely free of all PHI components.**

- [Gross image and annotations example](#)
- [Histologic image and annotations example](#)
- [Radiological image and annotations example](#)

### **For ALL representative images:**

- **Two identical sets** of representative JPEG images are required for gross, histologic and radiologic images that best illustrate features of the disease.
- The **first image** should be a clean image. The caption should state only the imaging modality (and appropriate sequence, if applicable) for radiology images and the magnification and stain for histology images.
- The **second identical image** can be marked using the “Annotate” tool provided. Use the arrows, circles, etc., to indicate the significance of each image. Captions should describe the annotated findings.
- Captioning image examples provided on pages 2-3 below.

### **Gross and Histologic Pathology Images (JPEG format only):**

- **All Specimen numbers must be removed.** Two identical sets (one clean image and one annotated image) of representative JPEG images are required for all submitted gross and histologic images, illustrating the features of the radiologic studies. Please note the magnification and stain for the histology images.

### **Radiologic representative images (JPEG format only):**

- When possible, radiographs should have images from more than one plane. Two identical sets (one clean image and one annotated image) of representative JPEG images are required. US should include gray scale and Doppler. CT should have appropriate windows, or sometimes multiple windows for the same image slice. MRI should have at least one image depicting appropriate sequences such as T1, fluid-sensitive sequence (T2 or STIR), pre- and post-contrast, etc.

### **Literature References:**

**\*\*IMPORTANT\*\*** All references must be either peer-reviewed journals or textbooks. Websites should not be referenced unless they are registries for the diagnosis (e.g. Pleuropulmonary Blastoma Registry). Please use AMA format for reference citations.

\* Instructional Video <https://youtu.be/KJ2BlexKG1A>

\*Case summary write up example provide pages 4-7 below.

## Captioning Images Examples: Gross, Histologic, and Radiologic Representative Images

**\*\*Please take note of the provided video links for image redaction and annotation**

Redaction Methods <https://youtu.be/ZppzfskKARQ>








Instructional Video <https://youtu.be/KJ2BlexKG1A>

**GROSS SPECIMEN IMAGES**

**Instructions:**

- [Gross image and annotations example](#)
- Click on **Upload Gross Specimen Images tab**. Use the pop-up window to select one or more gross image(s) from the previous Upload Images page.
- Check the box to select one or more image(s).
- Click **Upload** to finish uploading the image(s) on to the case summary page.
- Click **Annotate** to create a duplicate image with arrows, markers, and a descriptive caption.
- You may use the edit or delete buttons for your convenience.

[Upload More Gross Specimen Images](#)

Thumbnail	Caption	AIRP Index	File Name	Actions
	Cross section of the left kidney gross specimen.	G1	<a href="#">AIRP_15035034_gross ...</a>	  
	Cross section of the left kidney gross specimen. There is thinning of the renal cortex (yellow arrow and bar). There are dilated friable calyces (yellow circles).	G1.a	<a href="#">AIRP_150350...annotated</a>	 





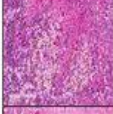


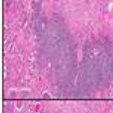






[Return to Top](#)

## HISTOLOGIC IMAGES















### Instructions:

- [Histologic image and annotations example](#)
- Click on **Upload Histologic Images tab**. Use the pop-up window to select one or more histo image(s) from the previous Upload Images page.
- Check the box to select one or more image(s).
- Click **Upload** to finish uploading the image(s) on to the case summary page.
- Click **Annotate** to create a duplicate image with arrows, markers, and a descriptive caption.
- You may use the edit or delete buttons for your convenience.

[Upload More Histologic Images](#)

Thumbnail	Caption	AIRP Index	File Name	Actions
	Histologic image or interstitium near renal pelvis in the excised left kidney. .	H5	<a href="#">foam-cells.jpg</a>	  
	Histologic image or interstitium near renal pelvis in the excised left kidney. There are clusterers of lipid-laden macrophages (yellow ovals) which is characteristic of xanthogranulomatous pyelonephritis.	H5.a	<a href="#">foam-cells ...annotated</a>	 
	Histologic image of renal cortex of the excised left kidney.	H2	<a href="#">kidney-2-56x.jpg</a>	  
	Histologic image of renal cortex of the excised left kidney. There is glomerulosclerosis (yellow circle). Few normal glomeruli (white circle) are seen on this image. There is diffuse infiltration of lymphocytes (blue oval).	H2.a	<a href="#">kidney-2-56...annotated</a>	 

## Radiologic Images


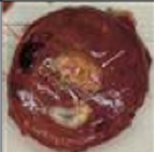
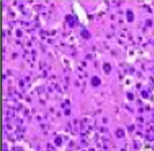
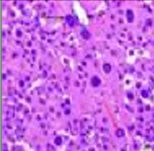


Image Type	Thumbnail	Caption	AIRP Index	File Name	Actions
CT with Contrast		Axial slice of CT abdomen and pelvis with contrast	CT2	<a href="#">axial-kidneys-1.jpg</a>	  
CT with Contrast		Axial slice of CT abdomen and pelvis with contrast at the level of the right renal hilum. There are bilateral renal calculi. There is enlargement of the left kidney, compared to the right side, with grossly dilated calyces (yellow arrows).	CT2.a	<a href="#">axial-kidne...annotated</a>	 
CT with Contrast		Coronal slice of CT abdomen and pelvis with contrast.	CT1	<a href="#">CT-coronal-1.jpg</a>	  
CT with Contrast		Coronal slice of CT abdomen and pelvis with contrast. There are large obstructing renal calculi (yellow arrow) and multiple bladder calculi (yellow oval). There are multiple cystic lesions in the spleen (red arrows). A cystic lesion in the inferior spleen abuts the superior pole of the left kidney where there is effacement of fat plane (green arrow).	CT1.a	<a href="#">CT-coronal-...annotated</a>	 

## Case Summary

### Clinical History

Patient is a 3-year-old white female with no significant past medical history who presented to her primary care provider with a 2-3 month history of increased muscle mass, facial/pubes hair, deepening voice, clitoromegaly, mild acne, and 5-6 months of rapid growth. Labs showed elevated for DHEA (dehydroepiandrosterone) 326.8 ng/mL (normal range 0.8-11.8 ng/mL) and total testosterone 5.37 ng/mL (normal range 0-0.2 ng/mL).

### Representative Images

Image Type	Thumbnail	Caption	File Name	Actions
Gross Specimen		Gross photograph of cut section of adrenal mass.	<a href="#">Gross 2.jpg</a>	<a href="#">Annotate</a> <a href="#">Edit</a> <a href="#">Delete</a>
Gross Specimen		Gross photograph shows extensive replacement of the adrenal gland by a red-brown nodular tumor with a focus of hemorrhage (red arrow) and necrosis (white arrow). Tumor weight was 525 g.	<a href="#">Gross 2_annotated.jpg</a>	<a href="#">Edit</a> <a href="#">Delete</a>
Histologic		H&E stained section of adrenal mass (original magnification, x400).	<a href="#">Histology High 300.jpg</a>	<a href="#">Annotate</a> <a href="#">Edit</a> <a href="#">Delete</a>
Histologic		Photomicrograph of adrenal mass shows large eosinophilic tumor cells with nuclear pleomorphism, hyperchromasia and irregular contours (black arrow). A few intranuclear "pseudoinclusions" are evident (red arrow) (original magnification, x400; H&E stain).	<a href="#">Histology H...annotated</a>	<a href="#">Edit</a> <a href="#">Delete</a>
Radiologic		Coronal contrast-enhanced CT image of the abdomen.	<a href="#">CT Coronal calcs nec...</a>	<a href="#">Annotate</a> <a href="#">Edit</a> <a href="#">Delete</a>
Radiologic		Right suprarenal mass causes inferior displacement of the right kidney. Central fluid attenuation within the mass is consistent with necrosis (white arrow). Scattered calcifications are present (white arrowhead).	<a href="#">CT Coronal ...annotated</a>	<a href="#">Edit</a> <a href="#">Delete</a>



Radiologic		Coronal fused PET/CT image of the abdomen.	<a href="#">PET coronal 300dpi.jpg</a>	<a href="#">Annotate</a> <a href="#">Edit</a> <a href="#">Delete</a>
Radiologic		Intensely FDG avid right suprarenal mass (white arrow). Central decreased FDG uptake is consistent with necrosis (red arrow).	<a href="#">PET coronal...annotated</a>	<a href="#">Edit</a> <a href="#">Delete</a>
Radiologic		Longitudinal ultrasound image of the right kidney.	<a href="#">US Long RK 300 dpi.jpg</a>	<a href="#">Annotate</a> <a href="#">Edit</a> <a href="#">Delete</a>
Radiologic		Homogeneous mass superior to the right kidney (white arrow).	<a href="#">US Long RK ...annotated</a>	<a href="#">Edit</a> <a href="#">Delete</a>
Radiologic		Longitudinal Doppler ultrasound image of the right suprarenal mass.	<a href="#">US Long RK Doppler ...</a>	<a href="#">Annotate</a> <a href="#">Edit</a> <a href="#">Delete</a>
Radiologic		Several foci of vascular flow identified within the heterogeneous right suprarenal mass (white arrow).	<a href="#">US Long RK...annotated</a>	<a href="#">Edit</a> <a href="#">Delete</a>
Radiologic		Transverse ultrasound image of the right suprarenal mass.	<a href="#">US Trans RK 300 dpi.jpg</a>	<a href="#">Annotate</a> <a href="#">Edit</a> <a href="#">Delete</a>
Radiologic		Hypoechoic area in the superior aspect of the right suprarenal mass (white circle) likely represents necrosis.	<a href="#">US Trans RK...annotated</a>	<a href="#">Edit</a> <a href="#">Delete</a>

## Diagnosis

Adrenal Cortical Neoplasm

## Differential Diagnosis

- 1: NEUROBLASTOMA
- 2: GANGLIONEUROBLASTOMA
- 3: PHEOCHROMOCYTOMA

## Demographics

Pediatric adrenal cortical neoplasms (ACN) have an estimated incidence of 25 cases per year in the United States. Unlike in adults, most (85%) ACN in children are hyperfunctioning. The most common clinical syndrome is virilization with elevated serum androgen (DHEA and testosterone) levels.

ACN demonstrate a bimodal age distribution, with an "infantile" group (<5 years) and an "adolescent" group (10-16 years). There is a female predilection in the infantile group. ACN has an increased incidence in several syndromes: Beckwith-Wiedemann, hemihypertrophy, Carney complex, multiple endocrine neoplasia type 1 (MEN-1), and neurofibromatosis type 1 (NF1). ACN in the infantile group, but not in adolescents or adults, has a high association with mutations of the p53 tumor suppressor gene. Li Fraumeni syndrome is a familial cancer syndrome caused by a germline mutation of p53. Affected patients are predisposed to many tumors including infantile ACN.

## Imaging Characteristics

On US, smaller neoplasms tend to demonstrate homogeneous hypoechogenicity or hyperechogenicity. Larger neoplasms have a more heterogeneous appearance. Central hypoechoic areas are consistent with necrosis. A hyperechoic rim can be seen. Color Doppler can be utilized in the evaluation of IVC invasion.

On CT, small tumors are homogeneous and large tumors are heterogeneous and displace adjacent structures. They frequently demonstrate central necrosis, calcifications, and peripheral enhancement. They may be surrounded by a thick enhancing rim. The findings of intracytoplasmic lipid and rapid contrast washout, which are so important in adults, are not useful in children. The reason is that most ACN in children are hyperfunctioning and surgical resection is indicated to cure the endocrine syndrome.

On MRI, large ACN demonstrate heterogeneous signal. High T1-weighted signal intensity compared to liver parenchyma suggests hemorrhage within the tumor. Internal high T2-weighted signal intensity indicates necrosis and hemorrhage. Contrast-enhanced MRI can more accurately detect invasion into vascular structures than contrast-enhanced CT. As ACN are metabolically active, Positron Emission Tomography (PET) is sensitive in the detection of distant metastatic disease.

## Differential Diagnosis Discussion

The main differential considerations for an adrenal mass in a child are neuroblastoma and ganglioneuroblastoma, which are much more common. These tumors are distinguished on the basis of the laboratory data, but an imaging feature that favors ACN over neuroblastoma is venous invasion. Findings that favor neuroblastoma include tumor crossing the midline, surrounding rather than invading vessels, and extension into neural foramina. Both ACN and neuroblastoma may demonstrate cystic change/necrosis and calcifications. Pheochromocytomas are tumors of the adrenal medulla which may occur in older children. Imaging features are similar to those of ACN but these tumors can be differentiated on the basis of laboratory analysis. Pheochromocytomas secrete catecholamines, such as epinephrine and norepinephrine. Metanephrines are catecholamine metabolites, which are also increased in pheochromocytomas.

## Pathology

ACN, comprising adrenocortical adenomas and carcinomas, typically present as unilateral, well-demarcated, small to large solid cortical masses that can be pink, tan, yellow or brown in color. The cut surface is homogeneous, nodular or lobulated with focal areas of necrosis, hemorrhage, or calcification.

The histologic appearance varies according to lipid content; lipid-depleted cells are small with eosinophilic cytoplasm, whereas lipid-rich cells are large and vacuolated. Tumor cells vary in appearance from uniform cells without mitoses to cells characterized by nuclear pleomorphism, hyperchromasia and numerous mitotic figures. Multinucleation and nuclear "pseudoinclusions" are occasionally evident. These represent invaginations of cytoplasm and they are not specific for ACN, as they may be seen in other tumors, including primary thyroid carcinoma and melanoma. This finding is not related to tumor size or function. Growth patterns may be alveolar, trabecular, or diffuse and infiltrative.

Unlike in adults, ACN in young children cannot be classified as benign or malignant on the basis of histologic evaluation alone. The only reliable predictor of biologic behavior is the finding of vascular invasion or metastases. For this reason, the term adrenal cortical neoplasm is preferred over adenoma or carcinoma.

## Treatment

Definitive treatment at all stages is surgical en bloc resection with adjacent invaded organs. If not possible to remove the entire mass, maximal tumor debulking is indicated to decrease the amount of hormone secreting tissue and diminish mass effect. Chemotherapy relies on the use of mitotane in both primary and adjuvant therapy, as well as in metastatic and recurrent disease. Radiation therapy is indicated in patients with a high risk for local recurrence, advanced local disease, and incomplete resection.

## Prognosis

Factors that have been associated with poor prognosis are tumor size greater than 10.5 cm, tumor mass greater than 400 g, extension to perirenal soft tissues or adjacent organs, IVC invasion, and metastases. ACN in the infantile group have a favorable prognosis, even with extensive disease.

## References

- 1 Young WF. Conventional imaging in adrenocortical carcinoma: update and perspectives. *Horm Cancer*. 2011 Dec;2(6):341-7.
- 2 Bharwani N, et al. Adrenocortical carcinoma: the range of appearances on CT and MRI. *Am J Roentgenol*. 2011 Jun;196(6):W706-14.
- 3 Chung EM, et al. From the radiologic pathology archives: precocious puberty: radiologic-pathologic correlation. *RadioGraphics* 2012; 32(7):2071-2099
- 4 Wieneke JA, et al. Adrenal cortical neoplasms in the pediatric population: a clinicopathologic and immunophenotypic analysis of 83 patients. *Am J Surg Pathol*. 2003;27(7):867-81.