

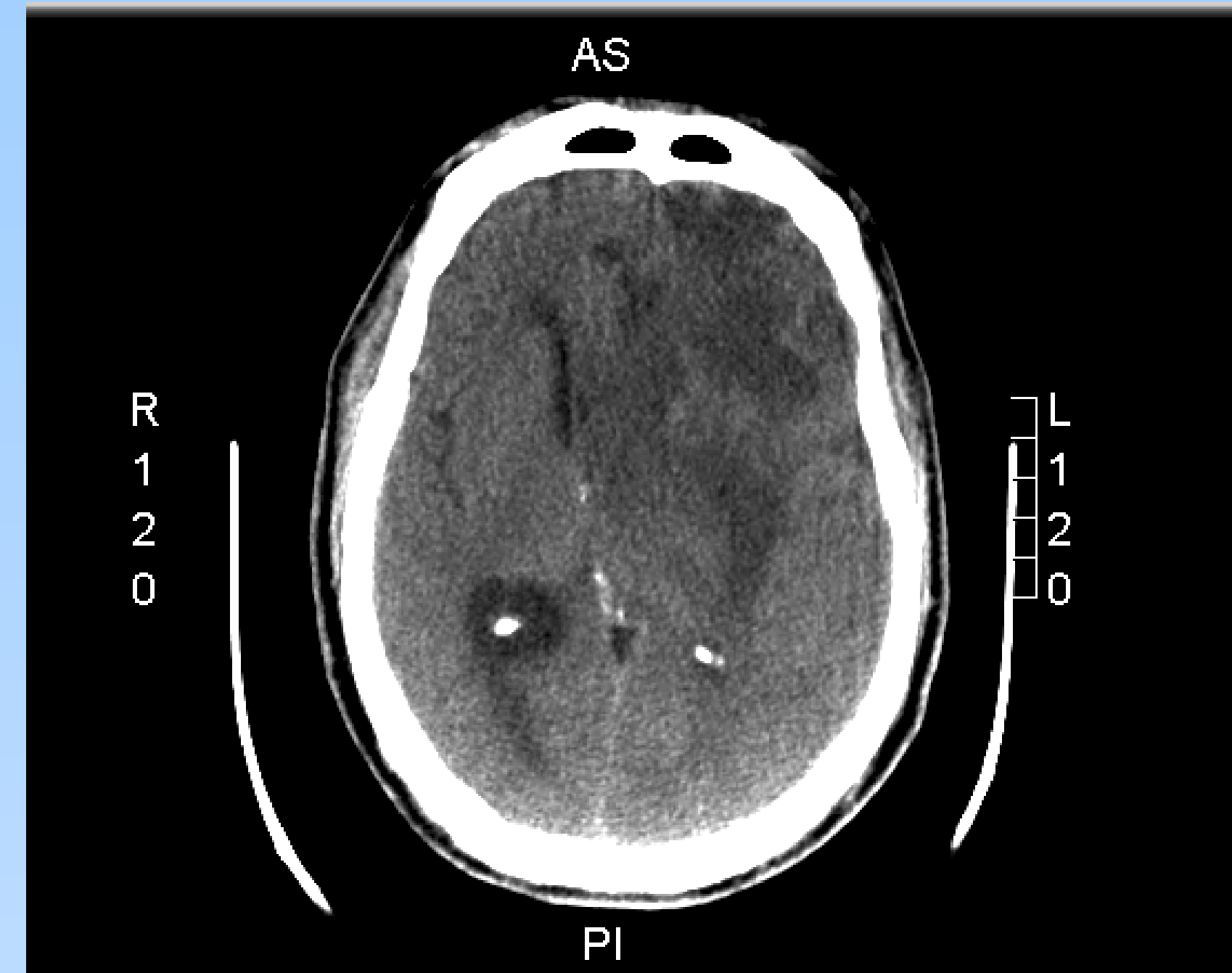
## Introduction

- Chondrosarcomas consist of a rare group of intracranial malignancies that are aggressive in nature.
- Mesenchymal chondrosarcomas represent the most malignant subtypes of these tumors, and can be usually be mistaken for other primary brain malignancies.
- I present a patient who presented with neurologic abnormalities, who was eventually found to have this extremely rare tumor, of which approximately 40 cases have been reported in the English-language literature.

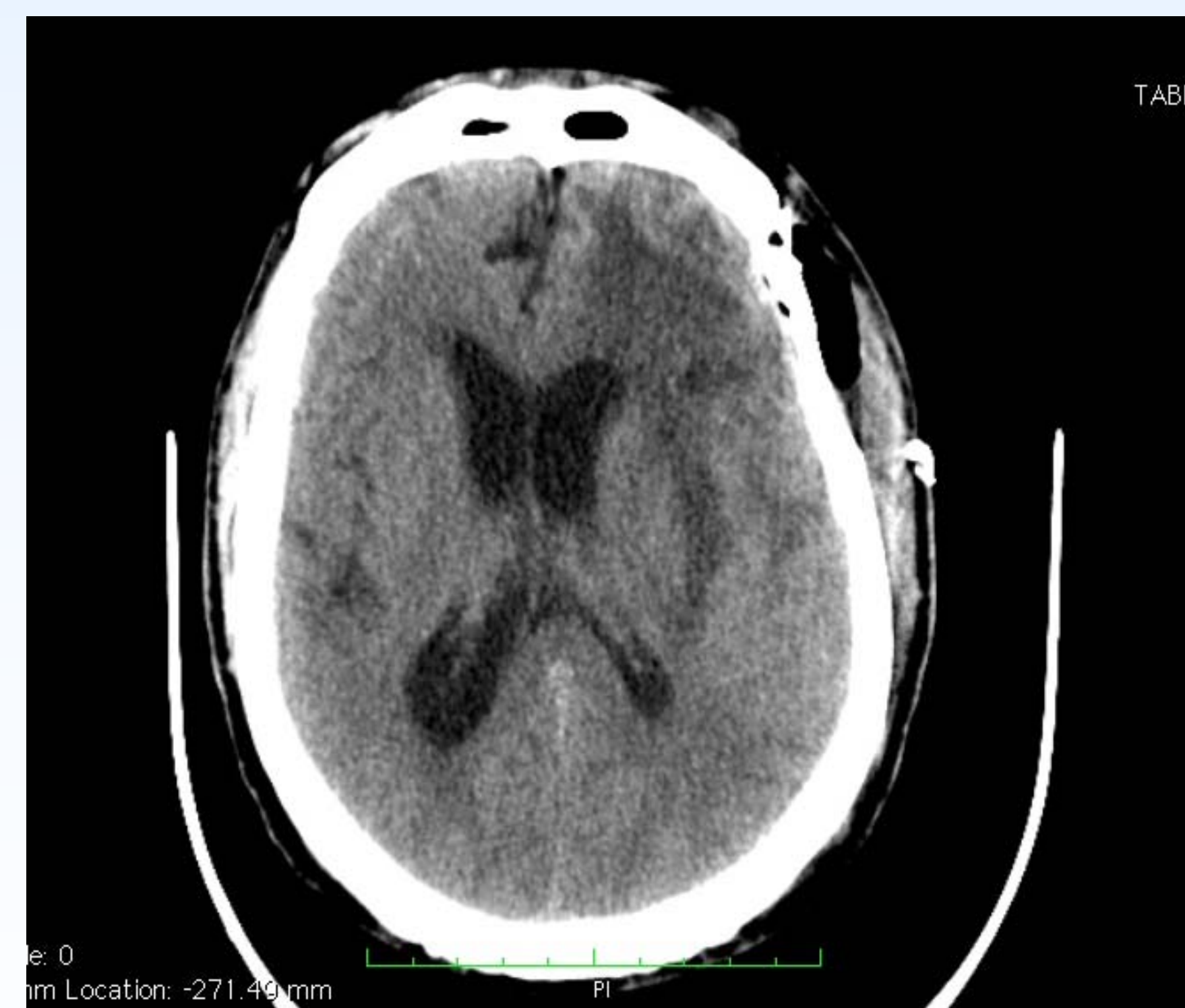
## Case Summary

- A 62 year old Caucasian male with history of substance abuse and hepatitis C infection was brought to the Emergency Room after being found down in his home and having two episodes of hematemesis.
- Although he was alert and oriented to name and month, he was not aware of his surroundings and why he was in the hospital.
- A neurologic exam was significant for equal pupils bilaterally but his left pupil was nonreactive to light, with a slight disconjugate gaze. Fundoscopic examination showed left sided papilledema. His extraocular muscles were intact but his left eye was unable to perform an upward gaze; ptosis was also noted in the left eye but no facial asymmetry. The patient also demonstrated left hemineglect but equal 5/5 strength in upper and lower extremities.
- An emergent CT study obtained showed a left frontal mass with vasogenic edema and midline shift. Neurosurgery was consulted and began following the patient.
- Several hours after admission the patient's neurologic exam and mental status began to decline and the patient was given mannitol, corticosteroids, and phenytoin, and his condition stabilized.
- An MRI confirmed the presence of a large brain tumor, and the patient underwent a left craniotomy with residual tumor in the area of the internal carotid and middle cerebral artery. The patient also underwent radiation therapy.
- However, postoperatively the patient's mental status declined due to hydrocephalus that developed later in the hospital course. Serial therapeutic lumbar punctures did not improve his mental state, and it was decided that placing a ventriculoperitoneal shunt would not improve his mental state or mortality.
- The patient also developed aspiration pneumonia and required prolonged intubation, and eventually expired 2 months after admission.

## Results

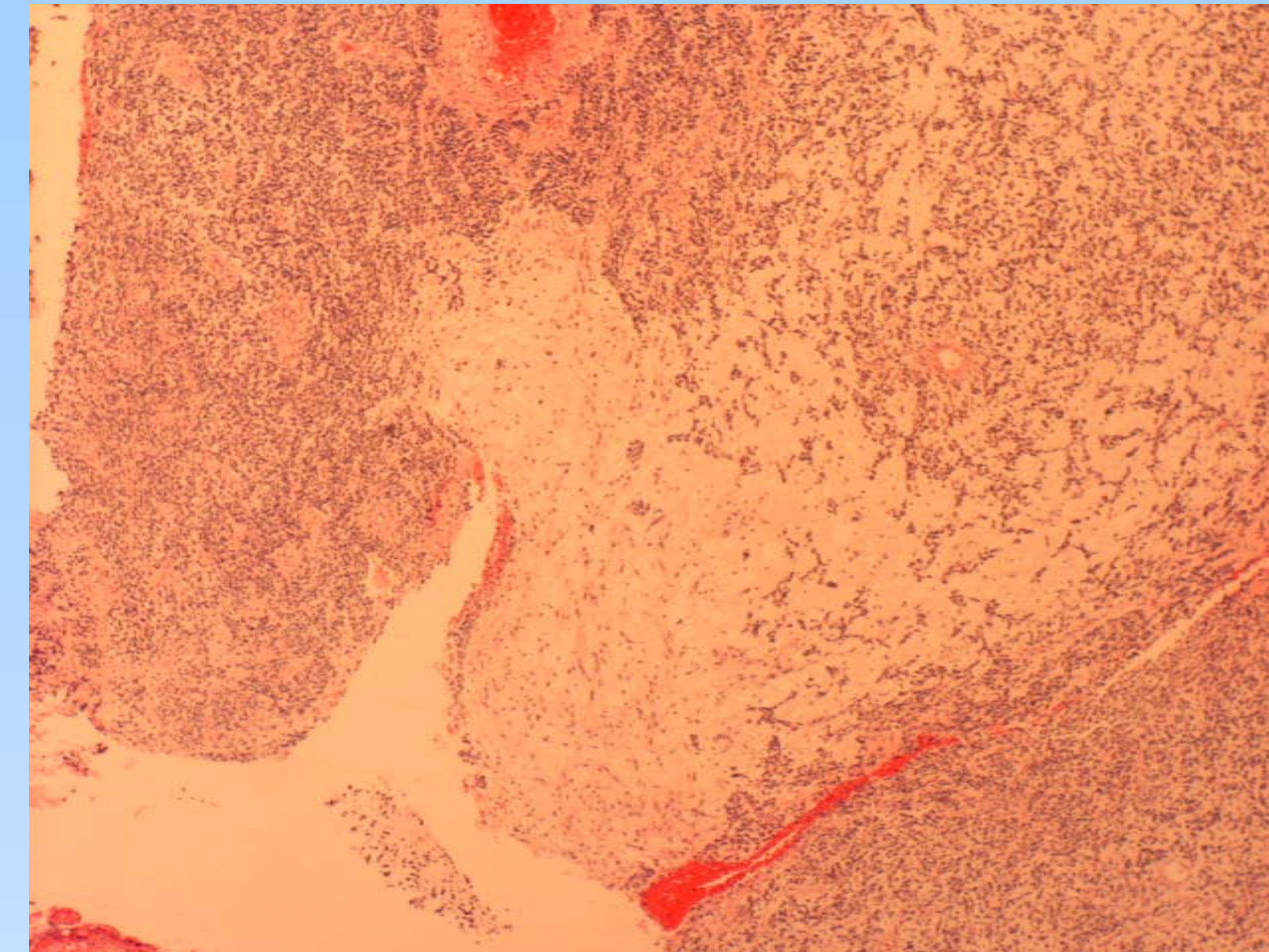


- A Computed Tomography study without intravenous contrast was obtained in the Emergency Room.
- In this cross section, there is a left frontal mass with a large area of vasogenic edema, as well as hydrocephalus seen in right ventricles with left uncus herniation. Left ventricles are not well visualized and there is midline shift to the right.
- A magnetic resonance imaging study with gadolinium administration was performed on the patient prior to biopsy.
- A large, approximately 5.0 X 5.6 cm intraaxial mass is located in the left frontal lobe with irregular thick enhancing walls with significant surrounding edema, mass effect and midline shift.
- Mild to moderate dilatation of right lateral ventricle and temporal horn was seen, consistent with moderate degree of hydrocephalus
- Enhancement is also seen along anterior aspect of medial portion of the left temporal lobe, suggesting involvement of superficial cortex or leptomeninges
- Findings are suggestive of aggressive primary brain tumor

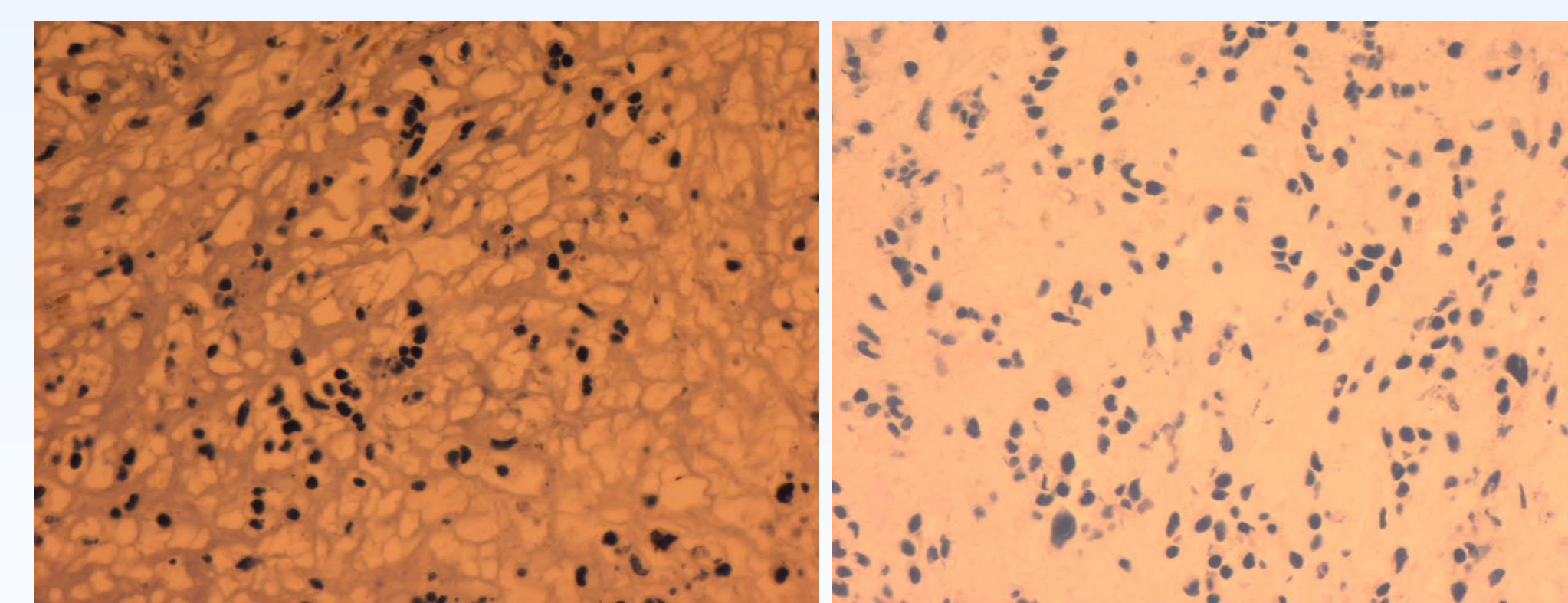


- This is a computed tomography without IV contrast study which was performed status-post craniotomy, showing improvement of mass effect and mild distention of the temporal horn of the right lateral ventricle.
- A follow up MRI showed peripheral nodular enhancement and seeding of tumor along the left tentorium and superior left cerebellum.

## Results

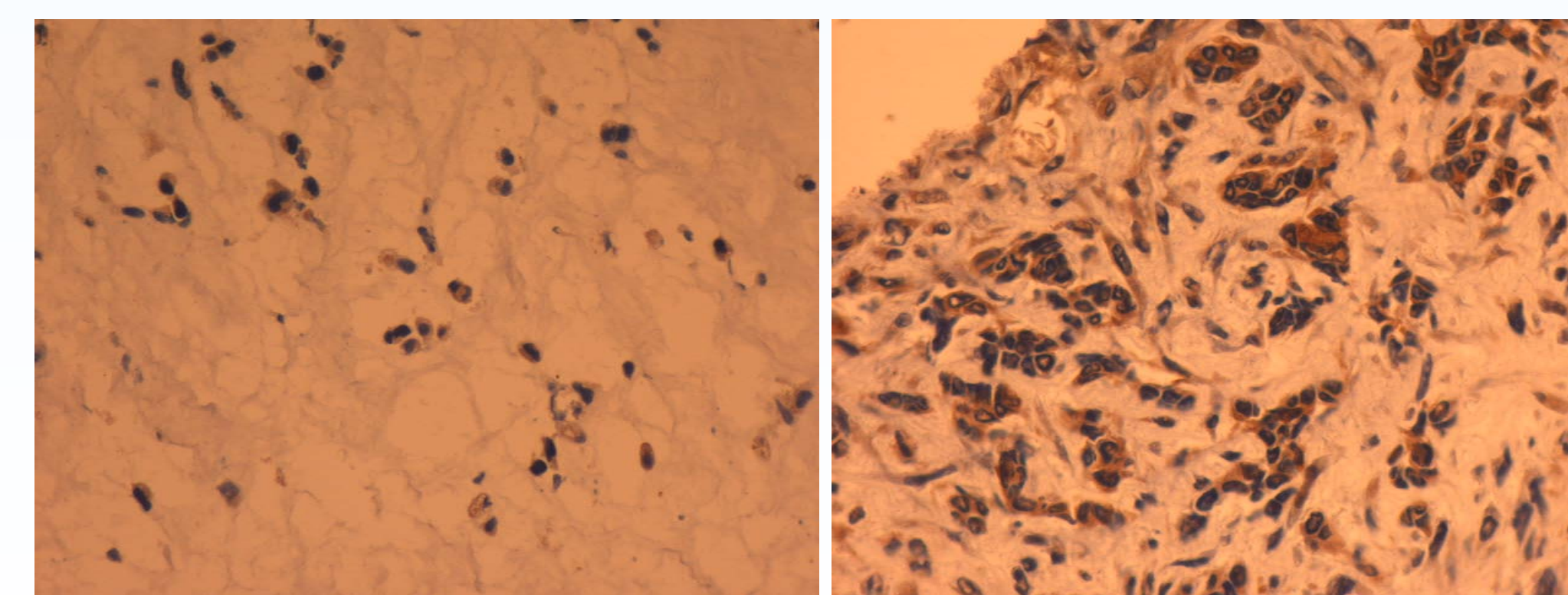


- Histopathological examination is shown above.
- Light microscope examination shows 2 distinct patterns
  - 1<sup>st</sup> pattern shows hypercellular containing tumor cells showing vesicular to hyperchromatic, oval to elongated nuclei, prominent nucleoli, nuclear molding, occasional nuclear grooving, and moderate to scant finely granular cytoplasm
  - 2<sup>nd</sup> pattern shows an area that is distinctly less cellular containing clusters of cells with lobulated, hyperchromatic nuclei and clear cytoplasm, and cells seen in background of extracellular matrix
- Electron microscopy shows majority of tumor cells have high nuclear:cytoplasmic ratio in hypercellular area. Some nuclei have complex lobation.
- Staining was negative for keratin (AE1/AE3), F8, EMA, positive for localization of S-100 and tau. CD34 positive in small vessel walls. Negative keratin and epithelial membrane antigen rule out chondroma and meningioma.



Keratin (AE1/AE3)

EMA



S-100

Tau

## Discussion

- Chondrosarcomas are a rare malignant disease constituting no more than 0.15% of the intracranial tumors., with three known classic subtypes: Myxoid, Mesenchymal, Classic.
- First case of intracranial mesenchymal chondrosarcoma described in 1962
- There are two subtypes of mesenchymal chondrosarcoma: one associated with muscle and soft tissues, and one associated with CNS tissue.
  - Geographically intracranial chondrosarcomas are supratentorial, usually located in frontoparietal region and attached to the meninges.
  - Histologically, the tumor has biphasic pattern of primitive small undifferentiated mesenchymal cells, among which are scattered islands of well-differentiated cartilage.
  - Radiographically, chondrosarcomas may resemble a hemangiopericytoma or a cartilage-containing meningioma, and it may be difficult to differentiate from a meningioma on MRI, often requiring biopsy.
- Most CNS extraskelatal mesenchymal chondrosarcomas reported in patients in their 2nd or 3rd decade of life, with slight female preponderance
- Soft tissue/muscle variant has mean age presentation of 43.9 years
- The mesenchymal subtype is the most Has propensity for local aggressiveness with recurrence, with metastases reported to occur in fewer than 20% of patients
- 5-year survival rate (including both skeletal and extraskelatal subtypes of the tumor) ranges from 42-68%, with 10-year survival rate ranges from 22-32%
- A literature search shows approximately 40 cases documented in English literature to date
- No randomized trials to determine efficacy of radiotherapy/chemotherapy
- Trials have evaluated the efficacy of combining proton radiation therapy, gamma knife radiotherapy along with cranial resection. A very small number of these patients studies had the mesenchymal subtype, and their prognosis tended to fare worse than classic subtype chondrosarcomas.
- Mainstay of treatment is aggressive surgical resection followed by radiation therapy, with close follow-up with still significant potential for local recurrence and distant metastasis.

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- ACKNOWLEDGEMENTS: Special thanks to Karen Kwan, MD, and Darryl Sue, MD for assistance on editing and feedback, and to the Departments of Pathology and Radiology for diagnostic images.