A Case of Angioimmunoblastic Lymphoma Presenting as Airway Obstruction

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Introduction

Angioimmunoblastic T cell lymphoma is a rare subset of Non-Hodgkin's Lymphoma. The incidence in the United States is 0.05 per 100,000 person years.([1]) It has a slight male predominance([2]) . It comprises 15-20% of the cases of peripheral T cell Lymphoma.([3]) The mean age at diagnosis is 69 years([4]). The prognosis is poor with a 5 year survival rate of 32%.([5]) The most common presentation is generalized lymphadenopathy. Non-Hodgkin lymphoma is known to involve Waldeyer's ring as the primary site of disease in 5% to 10% of cases.([6]) Here we present a case of advanced angio-immunoblastic lymphoma presenting as shortness of breath due to airway obstruction.

Case Report

An 83-year old female presented to the outpatient clinic with shortness of breath and right groin swelling. The difficulty in breathing was insidious in onset and progressed gradually to the point that patient had difficulties in activities of daily living. Patient also reported that she had developed noisy breathing since the last few months which worsened on lying down. Patient is a known case of Heart failure with preserved ejection fraction and long-standing hypertension, both these conditions were well controlled on medications. Physical examination revealed bilateral submental, supraclavicular, axillary lymphadenopathy, enlargement of bilateral tonsils and the adenoids along with right thyroid lobe enlargement. CBC revealed moderate anemia {Hb = 8.4/ hematocrit=25), normal WBC count (8700/ul) and thrombocytosis (platelet count 639000/ul). LFT revealed increased ALP (244; 30-130) and ALT (74; 6-29) levels. Serum albumin was low [2.7; reference: 3.6-5.1]. Haptoglobin levels were low (35; reference: 41-165). CT scan of the chest was remarkable for bulky mediastinal and axillary adenopathy. Imaging revealed bilateral pleural effusion with adjacent pulmonary consolidation. Brain CT showed nasopharyngeal mass 2*2 cm in size in the posterior wal of nasopharynx. Multiple hypodense lesions were seen in the liver. A provisional diagnosis of Non-Hodgkin's lymphoma was made. Patient was referred for biopsy of the right inguinal mass which revealed angioimmunoblastic variant of T cell lymphoma with EBV automated in situ hybridization of the tissue was positive. Ultrasound of the thyroid revealed right thyroid lobe enlargement with heterogenous echotexture. Anti TPO antibodies were in the normal range (<9IU/ml). The patient was started on MiniCHOP with Prednisone regimen. Subsequent improvement in breathing was evident on monthly follow up visits upto one year.

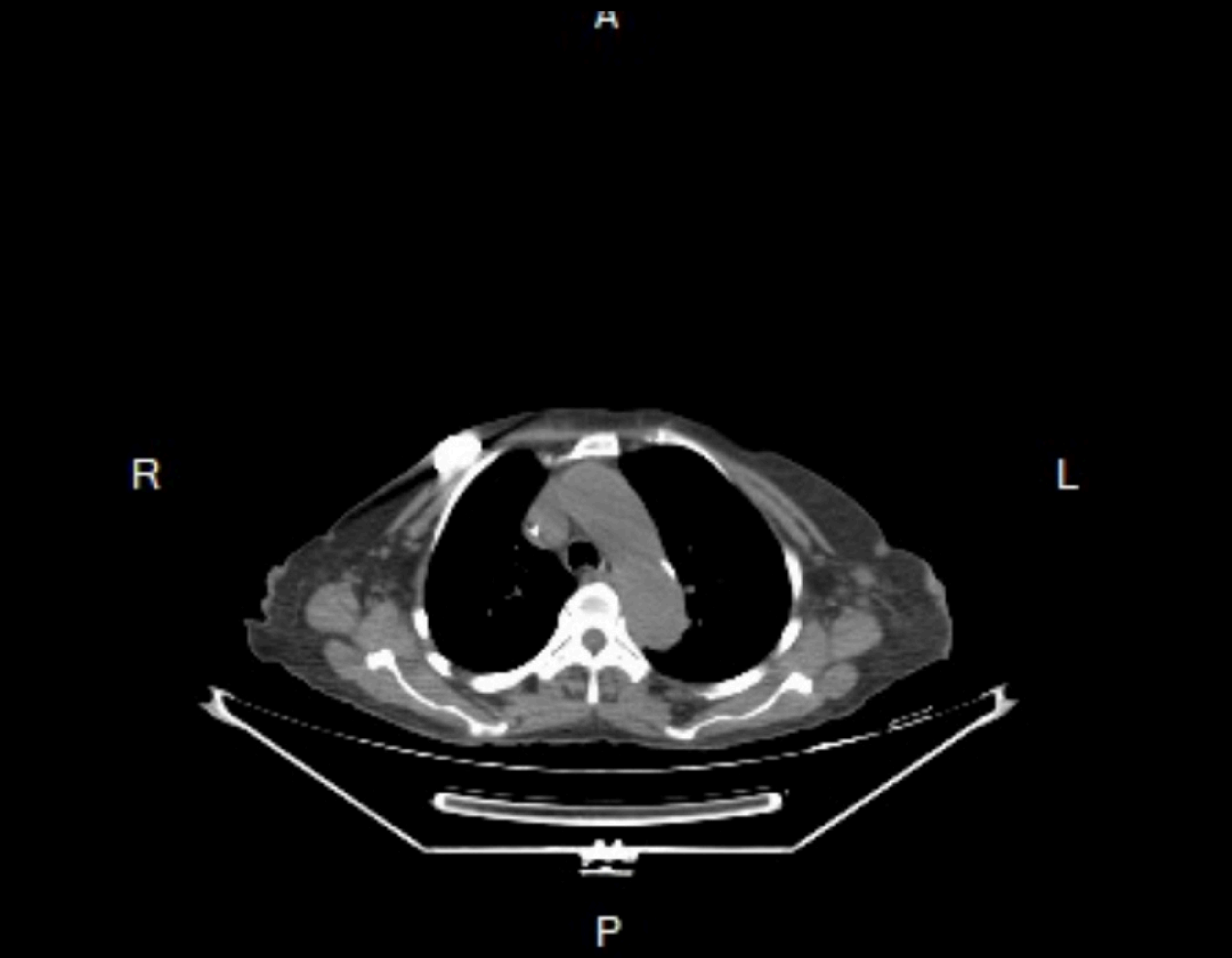
Discussion

Angioimmunoblastic T cell lymphoma is a rare disease of the elderly which usually presents as symmetrical generalized lymphadenopathy, B symptoms, rash and autoimmune cytopenias and autoimmune thyroid disease.([7]) The disease is commonly seen in Asians, Hispanics, and African Americans. Our patient presented with asymmetrical right inguinal lymphadenopathy. She also had involvement of the palatine tonsils, adenoids and the nasopharyngeal mucosal lymphatic tissue. Waldeyer ring involvement is rare in Non-Hodgkin lymphoma; particularly being large enough to cause symptomatic breathing difficulty. The nasopharyngeal mass possibly contributed to the noisy breathing. Asymmetrical thyroid lobe enlargement is a rather peculiar finding in Non-Hodgkin's lymphoma. The normal levels of TSH and anti TPO antibodies rule out autoimmune thyroid

disease, cause here could be metastatic. Thyroid enlargement along with mediastinal lymphadenopathy possibly contributed to the shortness of breath by narrowing the lower respiratory tract. The bilateral pleural effusion along with adjacent lung consolidation could be either due to metastases; it could also be due to worsening of patient's heart failure owing to exertion secondary to her respiratory distress. The patients lab results revealed thrombocytosis and normal leukocyte count which were a deviation from cytopenias([8]) seen in angioimmunoblastic T cell lymphoma. Total protein levels were low. Cold agglutinin antibodies test was positive which was consistent with the disease's association with autoimmunity. The patient had an increase in ALP and ALT levels. In the absence of history of alcohol intake, diabetes mellitus and obesity: these findings are suggestive of metastatic liver involvement.

Conclusion

Since the number of patients with the disease are limited, there is a dearth of large-scale studies evaluating the presenting symptoms of angioimmunoblastic lymphoma. The variations in laboratory abnormalities from routine findings also warrant further studies. The predominant involvement of nasopharyngeal mucosa leading to breathing difficulties in this case highlights the importance of having a high suspicion, since Angioimmunoblastic lymphoma can present as various unique clinical scenarios.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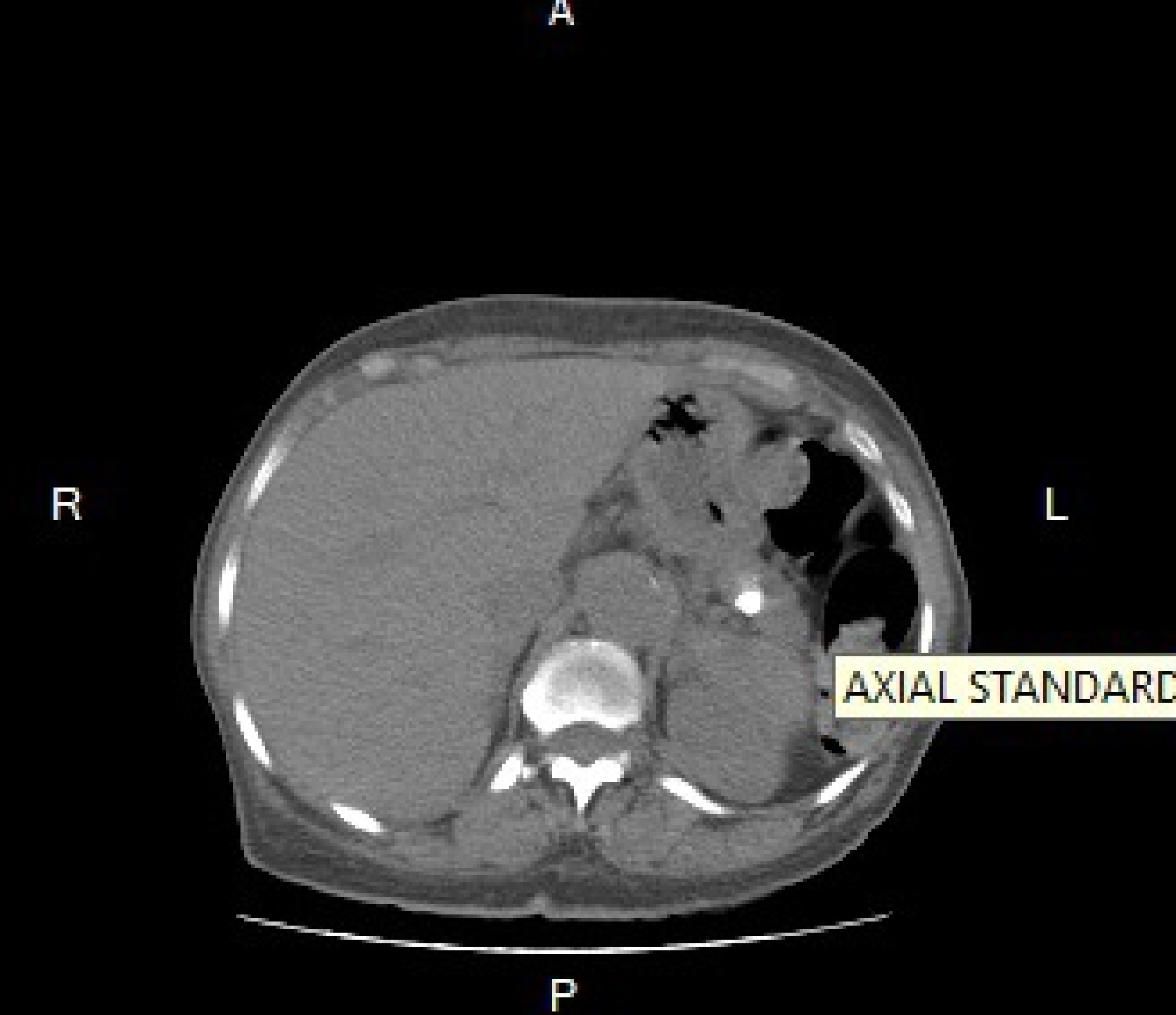
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