Squamous Cell Carcinoma of the renal calyceal system presenting with multiple overlapping features of Xanthogranulomatous pyelonephritis

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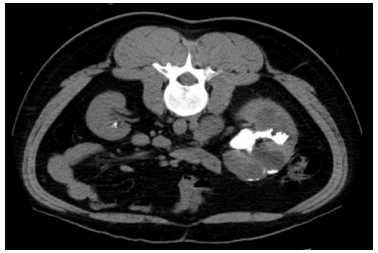
Introduction

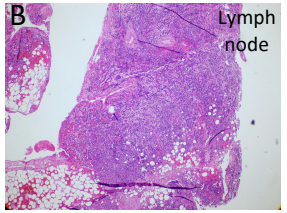
- Xanthogranulomatous pyelonephritis (XGP) is a chronic renal inflammatory disorder.
- Squamous cell carcinoma (SCC) arising from the renal calyceal system is a rare and aggressive cancer with poor prognosis due to its late-stage presentation and early metastases .
- XGP and SCC of the renal calyceal system have clinically similar presentations and it is important to differentiate between the two diseases.

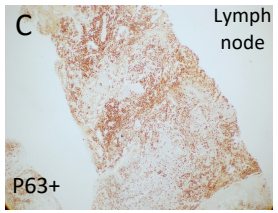
Case

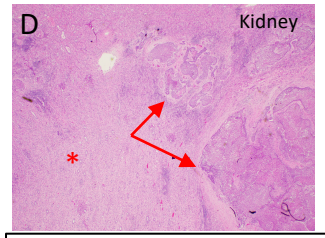
- A 37-year-old male with a history of recurrent nephrolithiasis and pyelonephritis presented with a 3-day history of right-sided flank pain and hematuria.
- Physical exam and lab workup was significant for right-sided CVA tenderness and UTI.
- CT imaging showed a large staghorn calculus in the right kidney.
- The leading diagnosis initially was XGP and UTI and patient was treated with antibiotics and discharged.
- Patient returned to hospital shortly with bilateral worsening flank pain.
- Treated with antibiotics and discharged after right nephrostomy stent placement.
- Patient called back to hospital after a repeat review of the CT scan showed IVC thrombus with retroperitoneal lymphadenopathy.
- Retroperitoneal lymph node (LN) biopsy showed extensive infiltration of the entire LN with neoplastic cells with disruption of normal architecture.

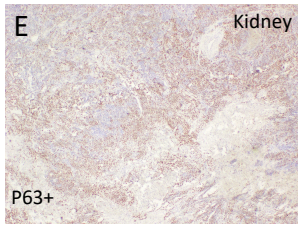
Radiological findings and Tissue Pathology

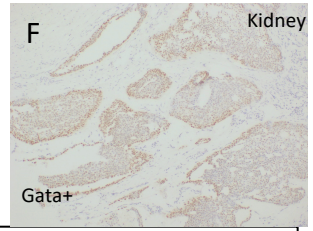












A. CT imaging: Large staghorn calculus and probable xanthogranulomatous pyelonephritis of the right kidney, adjacent retroperitoneal lymphadenopathy, and non-obstructing renal calculi in the left kidney **B.** Lymph node pathology showing disruption of normal architecture of lymph node and presence of large blue neoplastic cells. **C.** Lymph node IHC with p63 stain identifying cells of squamous epithelial origin. **D.** Kidney tissue pathology showing Infiltration of background kidney (red asterisk) by nests of neoplastic squamous cells (red arrows). **E.** IHC of kidney tissue with p63 identifying cells of squamous epithelial origin. **F.** IHC of kidney tissue with Gata identifying cells of urothelial origin.

Case (continued)

- The malignant cells stained p63+ on immunohistochemistry (IHC), indicating metastatic poorly differentiated SCC.
- Patient underwent radical right nephrectomy and IHC of the nephrectomy tissue showed tumor infiltration with neoplastic Gata3+ cells confirming the urothelial origin of the SCC.
- Repeat imaging showed disease progression with liver metastasis and despite chemotherapy, unfortunately patient died due to complications from metastatic cancer.

Discussion

- XGP is a destructive granulomatous process that can occasionally mimic other neoplastic and inflammatory processes of the kidney.
- In XGP, CT imaging and histology often reveal diffuse or focal disease with granulomatous inflammatory infiltrates.
- Chronic irritation in the setting of recurrent infections and inflammation can lead to urothelial dysplasia leading to renal SCC.
- Staghorn calculi are common contributors to the chronic inflammatory process that leads to cellular transformation.
- Thus, renal SCC must be considered in the differential diagnosis in patients with long-standing renal calculi, particularly staghorn calculi
- XGP results in high morbidity from chronic kidney dysfunction but the overall mortality from XGP is very low.
- In contrast, renal SCC has a 5-year survival rate <10% with a median survival time of 5 months.
- Early diagnosis and differentiation between these two conditions with very different prognoses can help targeted disease management.

Conclusion

Understanding rare and overlapping clinical presentations of an aggressive malignant condition (Renal SCC) with a chronic inflammatory process (XGP) can help achieve better patient outcomes.

References

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