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Opinion Article

Pancreatic Collision Tumors: A Rare Case of Extraosseous Ewing-like Sarcoma and Solid Pseudopapillary Tumor

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DESCRIPTION

A collision tumor refers to the unusual occurrence of two distinct histological neoplasms existing together in a single anatomical site. These tumors are exceedingly rare in the pancreas and their presence often indicates a poor prognosis for patients. The complexity of diagnosis and treatment of pancreatic collision tumors stems from the simultaneous presence of different types of tumors, each requiring unique management strategies. This article presents the first reported case of a pancreatic collision tumor consisting of an extraosseous Ewing-like sarcoma and a Solid Pseudopapillary Tumor (SPT) in a 43-year-old male.

Case Presentation

The patient, a 43-year-old male, was diagnosed incidentally with a pancreatic mass during routine imaging for an unrelated issue. The imaging findings were suggestive of a tumor involving the pancreas, but the nature of the lesion was unclear. Upon further investigation through biopsy and histopathological analysis, it was revealed that the lesion was a collision tumor, comprising two distinct types of tumors: An extraosseous Ewing-like sarcoma and a Solid Pseudopapillary Tumor (SPT).

The extraosseous Ewing-like sarcoma, a rare variant of the more common Ewing sarcoma, is typically characterized by small, round blue cells and exhibits aggressive growth patterns. While this sarcoma is known to primarily occur in bones, its occurrence outside the osseous structures is exceedingly uncommon. It is generally considered to have a better prognosis than other pancreatic malignancies but can still pose significant risks due to its potential for local invasion and metastasis.

The coexistence of these two histologically distinct tumors in a single pancreatic mass is a noteworthy anomaly. It confirms the need for heightened clinical awareness of collision tumors, especially in the pancreas, where they are infrequently diagnosed.

Diagnostic challenges

Diagnosing a collision tumor in the pancreas is a complex task, mainly due to the rarity of this phenomenon and the distinct histological features of the component tumors. Standard imaging techniques like Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) scans can often detect the presence of a mass, but they typically cannot differentiate between the specific types of tumors involved. In this case, the initial imaging findings raised suspicion of a pancreatic neoplasm, but the precise nature of the tumor was unclear.

Biopsy and histopathological examination are essential for accurate diagnosis. The use of immunohistochemically staining and molecular analysis enabled the identification of the Ewing-like sarcoma component, which exhibited the characteristic small round blue cell morphology and expression of markers such as CD99 and FLI-1. The SPT component was identified based on its distinct papillary architecture and positive staining for beta-catenin, a key marker for this tumor type. The simultaneous presence of both tumors in the same lesion made the diagnosis even more challenging and required careful differentiation between the two.

Therapeutic Challenges

Once diagnosed, the management of a pancreatic collision tumor involves addressing both tumor components and their respective treatment protocols. The therapeutic approach is complicated by the fact that one tumor (Ewing-like sarcoma) is highly aggressive, while the other (SPT) is typically indolent. The decision on how to prioritize treatment must be carefully considered.

This sarcoma, despite its rare occurrence in the pancreas, follows treatment protocols similar to those used for other soft tissue sarcomas, including multi-agent chemotherapy regimens and, where possible, surgical resection. Radiotherapy may also be indicated, especially in cases where surgical margins are positive or inoperable tumors are present.

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On the other hand, the treatment of SPT typically involves surgical resection alone, as these tumors are less aggressive and often have a favorable prognosis after removal. However, in the presence of a more aggressive tumor like the Ewing-like sarcoma, treatment protocols must be adapted to prioritize the more aggressive pathology to maximize patient outcomes.

A multidisciplinary approach is essential in the management of pancreatic collision tumors. Oncologists, surgeons, pathologists and radiologists must work together to develop a treatment plan that addresses both tumor components simultaneously. The priority is to treat the more aggressive tumor while ensuring that the less aggressive tumor is also appropriately managed. In this case, the management plan focused on the Ewing-like sarcoma's aggressive nature, with a treatment regimen designed to provide the best chance for survival.

CONCLUSION

Pancreatic collision tumors, while exceedingly rare, present significant diagnostic and therapeutic challenges. The case of a 43-year-old male with a collision tumor consisting of an extraosseous Ewing-like sarcoma and a solid pseudopapillary tumor highlights the complexity of managing such cases. Accurate and early diagnosis is important and a multidisciplinary approach to treatment is necessary for optimal outcomes. In particular, it is important to prioritize treatment for the more aggressive component of the tumor to ensure the best chance for patient survival. Given the rarity of these tumors, further studies and case documentation will be essential in improving our understanding of how to effectively manage pancreatic collision tumors and refine treatment protocols for these complex cases.