

# Intraabdominal Haemangiopericytoma Associated With Uncontrolled Diabetes Mellitus

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## Summary

Haemangiopericytoma (HPC) is a rare tumour with a predilection for the central nervous system. Though previously thought to originate from the meninges and ventricular walls, HPC's are currently accepted as distinct mesenchymal neoplasms unrelated to meningiomas. Haemangiopericytomas have been previously reported to be associated with the production of insulin like growth factor II (IGF – II) and hypoglycaemia. A case of a 61-year-old poorly controlled diabetic lady with a rare presentation of an abdominal haemangiopericytoma is discussed. A laparoscopic assisted resection of the massive lobulated tumour arising from the parietal peritoneum with dense attachment to the diaphragm and the liver was performed with an uncomplicated postoperative recovery. Control of the patient's diabetes improved dramatically following surgery and the rare association of hyperglycaemia and HPC, which has not been previously described, is elaborated upon in this report.

**Key Words:** *Hemangiopericytoma, Vascular tumors, Hyperglycaemia, Laparoscopic*

## Introduction

Hemangiopericytoma is an uncommon tumor that was first documented by Stout and Murray in 1942. It was not until 1949 when Stout reported on 25 additional cases that the tumor received widespread recognition.

Haemangiopericytomas are vascular tumors, thought to originate from the pericytes of Zimmermam which are unique cells founds spiraling around the outside of blood capillaries and post-capillary venules. This pathological entity is usually associated with an uncertain malignant potential due to its infrequent occurrence and unpredictable biological behavior. Histological confusion with other soft tissue tumors such as solitary fibrous tumor and synovial sarcoma is not uncommon due to the vascular nature of all the above-mentioned tumours. Modern diagnostic tools such as immunohistochemistry, electron microscopy with molecular and cytogenetic studies have however

been useful in differentiating HPC from other glomus tumours.

We illustrate the case of an elderly lady with an intraabdominal HPC that was associated with poorly controlled diabetes mellitus, contradicting the more commonly reported association of this tumour with hypoglycaemia.

## Case Report

A 61-year-old lady presented complaining of chronic intermittent right hypochondriac pain for 3 years with an associated mass over the same region. She had a background of non-insulin dependent diabetes mellitus and was hepatitis C positive, having undergone a total abdominal hysterectomy in the past. Abdominal examination revealed a mass in the right flank and right upper quadrant, measuring 15 X 10 cm that was semi-

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## CASE REPORT

mobile. Haematological investigations including serum biochemistry were normal apart from an elevated blood glucose. Her serum glucose remained erratically high during her hospitalization requiring high dose insulin therapy.

An abdominal computed tomography (Figure 1) revealed a large mass adjacent to and compressing the right lobe of the liver in the right upper quadrant. An ultrasound-guided biopsy of the mass favoured a diagnosis of a small cell tumor most likely neuroendocrine in origin. Surgical intervention in the form of a laparoscopic assisted resection was performed revealing a large lobulated tumor arising from the parietal peritoneum with dense attachment to diaphragm and the liver. The mass was resected utilizing an ultrasonic scalpel to minimize blood loss and the patient made a subsequent uncomplicated post-op recovery. The patient's diabetes improved dramatically following surgery without requiring insulin or oral hypoglycaemic agents. Histological analysis of the resected specimen confirmed a diagnosis of HPC.

### Discussion

HPC is a rare tumor that has been described as a protean tumor and while this is true with respect to its clinical presentation and anatomic distribution, its microscopic pattern is specific and reproducible. It is a neoplasm that is usually benign even though it has a definite malignant counterpart.

HPC is uncommon in children occurring primarily in adults with a peak incidence of 20-69 years with no racial or sexual preponderance. Pediatric cases account for less than 10% of all HPC's<sup>1</sup> in this age group. HPC may occur anywhere in the body, the most common anatomic location being the lower extremities, pelvis, retroperitoneum and the head and neck region<sup>2</sup>. It is usually a deep soft tissue mass found in muscle tissue with an insidious growth pattern. Other sites where HPC can occur include the trunk, upper extremities, orbit, intracranial region and the uterus and spleen. Dermal and subcutaneous lesions are much less common.

Clinical presentation of HPC's is non-specific with pain being a late symptom. Patients may complain of an associated enlarging mass. Symptoms however do vary depending on the site of disease. Most tumors present as a slowly growing mass that may cause intestinal or urinary symptoms in the abdomen. There have been numerous reported cases associated with hypoglycemia

due to secretion of insulin-like growth factor<sup>3</sup>, in contradistinction to our patient. Non-islet cell tumour of hypoglycaemia (NICTH) is a terminology used to describe haemangiopericytoma associated with hypoglycaemia. The mechanism that causes this metabolic abnormality is unclear even though raised blood levels of insulin like growth factor II (IGF-II) which has receptor binding abilities and a low protein binding affinity may theoretically result in increased free-type IGF-II in blood to express its biological activities. The suppression of hepatic gluconeogenesis with increased peripheral glucose uptake is also reported to cause hypoglycaemia in this tumour.

It is possible that there exists as yet an unidentified glucagon type growth factor that renders the patient diabetic in a small group of patients with HPC's. This may well explain the clinical manifestation of poorly controlled diabetes mellitus in our patient that subsequently normalized following surgery. We have found no similar documentation of an association between hypoglycaemia and HPC's on reviewing the literature. Radiographic findings of HPC's are not specific and the tumours usually consist of a well-circumscribed, radiopaque soft tissue mass that often displaces neighboring structures. Cystic changes are common, but calcification is rare and usually seen in large tumors of long duration.

Two distinct clinical entities have been described: the adult type which occurs in adults and children older than 1 year, and the infantile type occurring in the first year of life. Most infantile HPCs are considered congenital representing about one third of all paediatric HPC<sup>4</sup>. Prognosis is usually worse in the adult type. In a study of 106 HPC cases with available follow-up information on 93 patients, the reported 10-year-survival rate was 70%<sup>1</sup>. More recently, Espat *et al.*<sup>5</sup> reported an 86% five year overall survival rate. The authors however utilised a multimodal pathological review to exclude other soft tissue tumors that histologically mimicked HPC's<sup>5</sup>.

From a histological perspective, distinction between low-grade and high-grade lesions are difficult. Soft tissue hemangiopericytoma is a controversial pathologic entity. The relative non-specificity of the characteristic branching capillary pattern and the cytological features of the constituent cells, in addition to the lack of a distinct immunohistochemical staining profile, has resulted in uncertainty and absence of consensus regarding this subgroup of tumors<sup>1</sup>.

A review of the literature in adults suggests that the best mode of treatment for HPC's is resection with wide surgical margins. In Espot's report, patients undergoing complete tumor resection showed a 100% survival rate at five years<sup>5</sup>. Pre-operative ligation of afferent vessels or vascular embolization might help reduce the menace of operative hemorrhage<sup>2</sup>. In our case the use of a harmonic (ultrasonic) scalpel was successful in limiting blood loss. Both chemotherapy and radiotherapy seem effective and are recommended in all patients with incomplete resection or in the presence of a large locally invasive tumour<sup>2</sup>. Surgery and postoperative radiotherapy with 50 Gy or more has been suggested to significantly improve local control compared to surgery alone<sup>2</sup>. Local and distant relapses after prolonged disease-free interval have been reported<sup>5</sup>, suggesting a mandatory long-term follow-up<sup>4</sup>.

## Conclusion

HPC has a variable but predictable presentation and behaviour. Conventional malignant HPC is capable of both local recurrence and distant metastases, but has low disease-associated mortality. Due to the rarity of these tumors, there is however insufficient evidence to confirm the role of adjuvant chemotherapy in the management of this condition. The association of HPC with diabetes mellitus requires further elucidation, in particular the existence of as yet unidentified factors giving rise to hyperglycaemia. Utilising a laparoscopic assisted approach in resecting the tumour was helpful in procuring an effective outcome and early hospital discharge with no postoperative morbidity in this case.

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