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ABSTRACTS – POSTERS

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Retrospective analysis of patients with abdominal paragangliomas

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Background

Paragangliomas are rare chromaffin-cell tumours located at extra-adrenal sites along the sympathetic or parasympathetic nervous system. The main treatment option is surgical resection.

Up to 35% of abdominal paragangliomas are malignant, especially if associated with a SDHB (succinate dehydrogenase) gene mutation. The definition of malignancy in paragangliomas is the presence of malignancy with no other clear histological markers to predict the potential of recurrence or metastasis.

Aims

To report on the clinical and histological features as well as the outcome in patients with abdominal paragangliomas.

Methods

A retrospective analysis of the records of all patients diagnosed with abdominal paragangliomas between August 1998 and August 2008 was carried out.

Results

A total of 16 patients were operated on with curative intent for abdominal paraganglioma between 1998 and 2008. The mean age was 43 years (range 16- 71) at time of operation and sex distribution was similar with 9 female and 7 male patients.

Out of the 16 in total, 2 patients had a family history of pheochromocytoma, with 1 of these being a known carrier of the SDHB gene mutation. 1 patient was known to have MEN2 and 3 patients were diagnosed with a SDHB gene mutation following diagnosis of their paraganglioma

8 (50%) of patients presented with hypertension and 6 presented following investigation for back or abdominal pain. The patient with MEN 2 was diagnosed during routine follow up investigations and a further patient with family history was diagnosed on routine screening.

The most common site of disease was in the region of the celiac axis (10/16)

There was 1 post operative death. Apart from 1 patient developing a lower respiratory tract infection, there were no other post operative complications, with only 2 patients requiring HDU admission for haemodynamic monitoring.

In total 12.5 % (2/16) of patients have had recurrent disease at 4 and 6 years respectively following resection. Both had evidence of spread into the surrounding tissue on histology, however 4 patients who also had evidence of invasion into the surrounding tissues have had no recurrence to date (4- 30months post resection).

Conclusion

Due to their rarity, there is still little published on the clinical course of patients with paraganglioma. In this, one of the larger case series on abdominal paragangliomas we have shown that clinical presentation and size of the tumour were unrelated to prognosis, however Paragangliomas with evidence of extra-capsular spread are more likely to recur or metastasis, but this can occur many years following diagnosis.