UKINETS bitesize guidance

Guidance for the management of NETs of the Appendix

PAGE 1 - MANAGEMENT ALGORITHM

Acute appendicitis

Appendix mass

Appendicectomy

Neuroendocrine tumour identified - refer to specialist Neuroendocrine MDT/ Centre

Well differentiated Neuroendocrine tumour

Goblet cell Carcinoid/ MANEC/ Adenocarcinoma:

Refer to relevant guidance

Key Pathological Features

- **Size**
- **Location**
- **Depth of invasion**
- **Presence and depth of mesoappendix invasion**
- **Serosal involvement**
- **Vascular/ Perineural invasion**
- **Grade (mitotic index, Ki67%)**
- **Completeness of excision**

ENETS TNM classification used for staging

NET < 1cm (pT1 TNM staging)

R0 resection

Discharge from follow up

NET > 1cm and < 2cm

Mesoappendix invasion > 3mm

or Angioinvasion or Grade 2

Absent (i.e.pT2)

Present

Normal

Imaging

Single CT or MRI abdo/pelvis

NET > 2cm (pT3 or pT4)

NET with R1 resection

Imaging

CT or MRI abdo/pelvis

Consider somatostatin receptor scintigraphy or SR-PET/CT

Enlarged locoregional lymph nodes

Normal

or

Enlarged locoregional lymph nodes

Distant metastatic disease

See guidance on advanced mid-gut NETs

Surgery

Right hemicolectomy and nodal assessment

Follow up

6-12 monthly clinical assessment

Annual serum chromogranins

Annual CT TAP

Duration of follow up: 8 years

For further notes, including references, please see the following pages…

UKI NETS

UK and Ireland Neuroendocrine Tumour Society

For more information, please visit our website: www.ukinets.org

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PAGE 2 - NOTES & REFERENCES

Neuroendocrine neoplasms of the appendix are mainly diagnosed incidentally following appendicectomy with a rate of 3-5/1000 appendicectomies

Simple appendicectomy is adequate treatment for tumours <1cm in diameter where complete resection has been achieved, which have an excellent prognosis.[1-3]

For tumours > 2cm in diameter, there is a greater risk of nodal involvement and right hemicolectomy is therefore recommended.[1-3]

For tumours measuring between 1 and 2cm in diameter, risk factors for nodal involvement include the presence of deep mesoappendix invasion (>3mm), angioinvasion, and Grade 2 histology. Right hemicolectomy is therefore recommended for these patients.[1-3]

A recent meta-analysis of appendiceal well differentiated neuroendocrine tumours in children confirmed a higher risk of lymph node metastases in children with tumours > 2cm in size. However there was no recurrence or mortality observed in children who were treated with simple appendicectomy even when there were pathological criteria indicating the need for a right hemicolectomy.[6]

The frequency, nature and duration of follow up is poorly evidence based, but current guidelines recommend 6-12 monthly clinical assessment, and annual imaging for at least 8 years.[4,5]

References

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4. Boudreaux JP, Klimstra DS, Hassam MM, et al. The NANETS consensus guideline for the diagnosis and management of neuroendocrine tumours. Well differentiated neuroendocrine tumours of the jejunum, ileum, appendix and cecum. Pancreas 2010; 39: 753-766
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