Session Plan 1 – Please complete the sections shown in italics

Session title	Nerve Tumours in the Upper Limb
Session code	PLS_05_151
Session description	This session will describe the pathology and clinical presentation of nerve tumours in the upper limb. Later, it will explore the principles of treatment for patients with such tumours.
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Senior author (if applicable)	Dean Boyce
Deadline for submission of first draft to e-LPRAS	Monday 12th December 2016

What next?

Please check that the details in this table are complete and correct.
If not, contact elpras.project@googlemail.com
If correct, proceed to the next slide (Session Plan 2).

Session Plan 2 – Please complete the sections shown in italics

Learning objective 1	Describe the pathology of nerve tumours in the upper limb
Learning objective 2	Describe the patterns of clinical presentation of nerve tumours in the upper limb
Learning objective 3	Explain the principles of treatment
Learning objective 4	N/A
Learning objective 5	N/A
Prerequisites	*Is there anything the student should have revised or read before studying this session?*
Related sessions	PLS_01_059_Peripheral Nerve Tumours PLS_01_060_Neurofibromatosis PLS_05_152_Management of Ganglion in the Hand PLS_05_153_Glomus Tumour in the Hand PLS_05_154_Giant Cell Tumour of the Tendon Sheath in the Upper Limb
Keywords	*Students will be able to search for your session based on key words.*

What next?

Please do not alter the learning objectives!
If you believe that they need to be modified please contact elpras.project@googlemail.com
Proceed to the next slide (Author Biography).

Author Biography – This slide needs to be completed by the Content Author.

First Author

Ernest Azzopardi is honorary senior clinical lecturer at Swansea University, UK and Specialist registrar at the Welsh Centre for Burns and Plastic Surgery. Link to online profile:

http://www.bapras.org.uk/professionals/research/academic-careers/academic-profiles/ernest-azzopardi

Senior Author

Dean E Boyce is honorary senior clinical lecturer at Swansea University and senior consultant at the Hand and Peripheral Nerve Unit, the Welsh Centre for Burns and Plastic Surgery

Link to online profile

Session Introduction – This slide needs to be completed by the Content Author

Hand tumours represent 15% of all soft tissue lesions presenting in the entire body. Peripheral nerve tumours in the upper limb represent 1-5% of all hand tumours.

Neuromas and schwannomas represent the commonest peripheral nerve tumours. Other nerve tumours in the upper limb comprise neurofibromas. Malignant nerve tumours in the upper limb include neurofibrosarcomata and malignant peripheral nerve sheet tumours (MPNST).

Useful algorithms to bear in mind for both clinical practice and exit exams are the following

A: What is it? How bad is it? What management?

B: Benign v Malignant and Primary v Secondary

C: Surgery vs other modalities

D: Resectable vs irresectable

NHS

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What next?

Please supply a short introduction to your session in the space provided.
Then proceed to the next slide (Content Page).

Content Page – The Content Author will use this slide many times for content.

Schwannoma

Incidence: Schwannomas are the commonest peripheral nerve tumours. They represent benign neoplasms of Schwann cell origin, from the peripheral nerve sheath.

Symptomatology: May arise as a direct effect of the tumour- classically a slow growing painless mass; and due to mass effect in a closed compartment. pain, paraesthesia/weakness.

Examination classically, reports a smooth surfaced and edged, mobile under overlying skin but fixed. Tinel's test may be positive.

Natural history: Malignant degeneration is uncommon.

Investigation: Schwannomas exhibit a classic onion-skin appearance on MRI.

Management: Most tumours are encapsulated and separable from the nerve of origin. Classically, schwannomas are well-defined, and can be extirpated by intraneural dissection, in which case recurrence is rare.

Image

clinical schwannoma

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Neurofibromata

Neurofibromata are rarer than neuromas

Pathology: Unlike Schwannomas, neurofibromata are not encapsulated and are intimately associated to nerve fascicles. They arise from the central nerve fascicles

Clinical presentation: soft, superficial lesion, smooth surface and edge, non-compressible. They can occur as dermal or plexiform. Plexiform neurofibromata can arise as nodular or diffuse (cf NF module). Seek stigmata of NF.

Indications for excision in the upper limb include functional limitation by virtue of size and location.

Risk of Malignant Transformation: For solitary neurofibromas the risk of malignant transformation is rare. However, NF does carry a risk of malignant transformation (10-15% of cases). Classically this is heralded by increased rate of growth and pain.

Investigation: MRI is the preferred mode of imaging, for diagnostic confirmation and mapping the extent of involvement of the associated nerve. Due to the possible functional significance, biopsy may be indicated to confirm the diagnosis, resectability and likely functional deficit

Image:

Clinical appearance

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The signal intensity of benign PNST's is fairly nonspecific, but, in contrast to MPNST's shows isointensity or slight hyperintensity to muscle on T1 modality. T2 weighted images show hyper intensity.

A target sign with hyperintense periphery and central hypointensity on T2 weighted is most commonly seen with neurofibroma, but occurs with other PNST.

Heterogeneity is more in keeping with MPNSTs.

The ''split fat'' sign is often seen around neurogenic tumours: neurovascular bundle is surrounded by fat so masses arising from this location maintain a rim of fat around them.

The ''fascicular'' sign describes small ring like structures on T2 weighted images corresponding to the fascicular bundles within the nerve.

Image:

MRI appearance

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Management: The key considerations here are the functional significance of the associated nerve (low v high) and the resectability of the Neurofibroma (need for excision and repair). In nerves of minor importance, excision and repair (example via direct repair or nerve cable grafts) may be reasonable.

Neurofibromas can sometimes be separated from individual fascicles without need for nerve excision, in larger nerves.

If the Neurofibroma is diffusely associated with a nerve of high functional significance then the residual deficit, likelihood of recovery and timing of recovery need to be considered on an individual bases.

Image:

MRI appearance

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Neuromas

Neuromas are benign lesions which occur in two varieties depending on the aetiology.

1. Neuroma in continuity

Spindle variety: chronic irritation in an intact nerve such as the lateral cutaneous n. of the thigh

Lateral variety: develops at a site of partial nerve division of following nerve repair

2: End –neuroma: often follows traumatic nerve division or amputation

Common sites for neuroma formation are

1. Palmar cutaneous branch of the ulnar nerve
2. Superficial branches of radial or radial digital nerve
3. Dorsal branch of ulnar

Clinical appearance of a neuroma

What next?

Duplicate this slide as many times as you need to create extra content pages.
When you have all of your content, proceed to the next slide (Key Points).

Content Page – The Content Author will use this slide many times for content.

Management for neuromata depends on the aetiology. Options include

Non-surgical: desensitisation exercises, transcutaneous electrical nerve stimulation, and medical management including carbamazepine. Referral to a pain specialist may be appropriate.

Surgical: several techniques have been described, broadly divided into two categories

A. resection and coagulation (chemical, laser, bipolar diathermy); ligation; crushing; capping; epineurial repair over the cut end

B. Burying: in nearby bone or muscle; implantation into another nerve (such as for bilateral digital amputation neuromas).

C: combined techniques such as “crush cap and flip” where the nerve is crushed proximally to induce neuropraxia, then capped, and then flipped over to be buried into deep tissues)

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Intra neural non-neural tumours

Intraneural non-neural tumours have been reported. This distinct minority includes intraneural lipoma, hemangioma, lipomatosis of nerves. There is a distinct association with macrodactyly.

Malignant peripheral nerve sheet tumours

Incidence: Malignant peripheral nerve sheath tumours (MPSNT) comprise 2-3% of malignant hand tumours, and are associated to local extension and metastasis. MPNST is associated to mutations in *TP53* mutations. It occurs frequently in NF1 (9-13 % lifetime risk). However It appears to be only increased in NF2 amongst those that have been irradiated.

Malignant Triton Tumour is an MPNST variant showing rhabdomyosarcomatous differentiation

Clinical symptomatology: As with other sarcomas, cardinal signs include

- Increase in size
- Size > 5 cm
- Deep tissue plane involvement
- Increased pain

Natural progression: MPNST usually metastasize in a haematogenous fashion, most commonly to the lungs

MPNST also presents with mass effect signs including irritability of the primary nerve. Clinical manifestations of this include signs of irritability (pain, tinel's sign) sensory, motor and vaso-trophic changes

Investigation of suspected MPNST follows sarcoma protocols, and as such necessitates discussion in a sarcoma MDT. Diagnostic biopsies are planned following potential resection incision lines, and imaging is performed by the MDT radiologist with an interest in sarcoma. Imaging options include Ultrasound, MRI and PET-CT.

Management.

Management within an MDT setting is mandatory. Modalities of treatment include surgery, and radiotherapy.

Surgery remains the primary treatment method, and may be performed in combination with radiotherapy. (CF: sarcoma section). Radiotherapy can have both a neoadjuvant or adjuvant role. Pre-surgery, this may result in tumour shrinkage. Post surgery, radiotherapy may be indicated in intermediate or high grade tumours, and those with close resection margins.

Amputation versus limb salvage: amputation is considered when critical structures need to be excised leaving a limb with poorer function then a prosthetic. Limb salvage requires a multimodal approach to achieve a low recurrence rate

Key Points – This slide needs to be completed by the Content Author

Achievements: By the end of this module, you should be able to describe the pathology of nerve tumours in the upper limb, patterns of presentation , and a logical method of determining treatment.

Key summaries: peripheral nerve tumours in the upper limb represent 1-5% of all hand tumours. Schwannomas are the commonest benign peripheral nerve tumours

Schwannomas are encapsulated and commonly easily resectable whereas neurofibromata are not encapsulated.

MPNST is rare and management should be planned within a sarcoma MDT setting. Surgery remains the mainstay of treatment with or without neoadjuvant / adjuvant radiotherapy

NHS

e-LfH

e-Learning for Healthcare

References

Gareth D, Evans R, Huson S, Birsh J. Malignant peripheral nerve sheath tumours in inherited disease

*Clinical Sarcoma Research*2012**2**:17

Hsu, Charles S., Vincent R. Hentz, and Jeffrey Yao. "Tumours of the hand."*The lancet oncology* 8.2 (2007): 157-166.

Hirose, Takanori, Bernd W. Scheithauer, and Toshiaki Sano. "Perineurial malignant peripheral nerve sheath tumor (MPNST): a clinicopathologic, immunohistochemical, and ultrastructural study of seven cases." The American journal of surgical pathology 22.11 (1998): 1368-1378.

Stucky, Chee-Chee H., et al. "Malignant peripheral nerve sheath tumors (MPNST): the Mayo Clinic experience." *Annals of surgical oncology* 19.3 (2012): 878-885.

Zaidman CM, Seelig MJ, Baker JC, Mackinnon SE, Pestronk A. Detection of peripheral nerve pathology Comparison of ultrasound and MRI. Neurology. 2013 Apr 30;80(18):1634-40.

What next?

Please enter your references in the space provided.
Then proceed to the next slide (Further Reading and Next Steps).

Further Reading and Next Steps – This slide needs to be completed by the Content Author

Zaidman, Craig M., et al. "Detection of peripheral nerve pathology Comparison of ultrasound and MRI." *Neurology* 80.18 (2013): 1634-1640.

Pekmezci, Melike, et al. "Morphologic and immunohistochemical features of malignant peripheral nerve sheath tumors and cellular schwannomas."*Modern Pathology* 28.2 (2015): 187-200.

Farid M, Demicco EG, Garcia R, Ahn L, Merola PR, Cioffi A, Maki RG. Malignant peripheral nerve sheath tumors. The oncologist. 2014 Feb 1;19(2):193-201.

Suggested next step activities:

If available in your area, attending the following clinical sessions may be useful.

1. Sarcoma MDT
2. Sarcoma screen clinic
3. Watch principles of sarcoma excision
4. Attend follow up sarcoma clinic

What next?

Please enter your references and next-step activity idea(s) in the spaces provided.
Then proceed to the next slide (Question Page).

Question Page – This slide needs to be completed by the Content Author.

Question.

A. Answer

B. Answer

C. Answer

D. Answer

E. Answer

Enter your feedback here – produce either: A) a general paragraph; or B) individual comments as shown in the example below.

A. Incorrect. Explanation.
B. Incorrect. Explanation
C. Correct. Explanation
etc

What next?

If you are happy to do so, duplicate this slide to create up to 4 multiple-choice questions. Then proceed to the next slide (Checklist).

Checklist – This slide needs to be completed by the Content Author

Success Criteria	Yes/No?
1. Have you included an author biography?	1.
2. Have you covered all of the learning objectives?	2.
3. Have you supplied images for each slide or made clear what is required?	3.
4. Have you given key points to summarise?	4.
5. Have you supplied references for further reading?	5.
6. Have you suggested at least one next step activity?	6.
7. Have you supplied MCQs (optional)?	7.
8. Has your senior author (if applicable) approved the content? If you have not passed FRCS(Plast) you must have a senior author.	8.

What next?

Review the points shown in the checklist to ensure that your content is ready for submission.
Once complete, save as 97/2000/XP.ppt and email to: elpras.project@googlemail.com.
THANK YOU VERY MUCH FOR YOUR HARD WORK!! WE WILL BE IN TOUCH SOON