

Minor Surgical Procedures Required for Establishing Diagnosis in Neuropathology in Children: A Brief Guide for Clinicians

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Abstract

In clinical neurological practice, histopathological diagnosis is often required for establishing definitive diagnosis of various neuromuscular disorders and neurodegenerative disorders like neuronal ceroid lipofuscinosis. To obtain the tissue specimen required for putting histopathological stains, various minor surgical procedures are often carried out by the clinicians. These include nerve biopsy, muscle biopsy and skin biopsy. A clinician needs to be aware of the ideal procedure, proper clinical indications, perioperative and postoperative care of patients undergoing these minor surgical procedures. This review article provides a brief review of the above three procedures in a concise manner. Muscle biopsy is usually carried out in children with suspected muscular dystrophies and myopathies, and the most common site is vastus lateralis. Similarly nerve biopsy is carried out in children with suspected hereditary and acquired peripheral neuropathies from sural nerve. Skin biopsy is usually carried out from axilla, in children with neuronal ceroid lipofuscinosis to demonstrate autofluorescent granules and fingerprint/curvilinear or rectilinear inclusions.

Key words: Muscle biopsy, Nerve biopsy, Skin biopsy, Children

Introduction

In clinical neurological practice, histopathological diagnosis is often required for establishing definitive diagnosis of various neuromuscular disorders and neurodegenerative disorders like neuronal ceroid lipofuscinosis. To obtain the tissue specimen required for putting histopathological stains, various minor surgical procedures are often carried out by the clinicians. These include nerve biopsy, muscle biopsy and skin biopsy. A clinician needs to be aware of the ideal procedure, proper clinical indications, perioperative and postoperative care of patients undergoing these minor surgical procedures. This review article provides a brief review of the above three procedures in a concise manner.

Muscle Biopsy

Indications:

Evaluation of various neuromuscular disorders such as suspected myopathies, muscular dystrophies, mitochondrial disorder [1].

Contraindications:

Any muscle which has been the site of the following should not be biopsied.

- Prior intramuscular Injection
- Previous biopsy
- Electromyography (EMG) studies

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- Prior trauma

These procedures result in artifact changes in the muscle in the form of inflammation and reactive changes which may produce confusion in the interpretation of the biopsy [1].

Prerequisites:

- Ensure that the child is fasting for at least 4 hours before the procedure in case of infants 3 hours of fasting is enough. However, if they need to wait further IVF should be started.
- A short admission for daycare should be made.
- Proper, written and informed consent should be taken from the parents. If parents are not available then whoever has brought the child for treatment needs to give consent on behalf of the child parents. The parents and guardians should be explained about the procedure, risk of sedation and possible complications and consent form should be signed.
- Chest radiograph- Look for enlarged heart with cardiomyopathy or pulmonary problems
- ECG in patients > 2 yrs and preferably echocardiography in patients < 2 yrs to look for the cardiomyopathic component in some structural and metabolic myopathies [2].
- Prior CPK and EMG should be done before doing muscle biopsy as CPK is expected to be elevated after muscle biopsy.
- The site of EMG needle insertion and muscle biopsy should be different to avoid artefactual changes on histological examination. Muscle biopsy may be done on Right side and EMG on the left to avoid sampling error.
- Ensure that in the treatment room sedation is ready Midazolam and Ketamine in labeled syringes with appropriate dilution, the muscle biopsy sterile set, blade, suture and the vial for transport of muscle biopsy sample duly obtained beforehand from the neuropathology laboratory [2].

Selection of muscle:

- **Muscular dystrophies:** A moderately affected muscle should be selected. A minimally involved muscle may not show definitive histological changes while a severely involved muscle would show changes of endstage fibrosis. Usually **Vastuslateralis** is the muscle which is preferred in this situation.
- **Acute inflammatory muscle disorders:** For conditions like dermatomyositis or polymyositis, most severely involved muscle is selected. A preceding MRI of the muscle helps in selection of the muscle to be biopsied.

Steps of Muscle biopsy:

- Decide the site of biopsy. Prepare the site with betadine and spirit as described earlier for other procedures. Drape the wound.
- An incision of 3-4 cm length is made in the skin over the

middle of the belly of the muscle in the direction of the fibers.

- The skin is retracted and the fascia is incised to expose the muscle fascicles.
- Hemostasis is achieved using pressure with gauze pieces.
- The direction of the fibers is determined and a cylinder of muscle, about 0.5 cm across, is carefully isolated from the main muscle bulk by blunt dissection using artery forceps.
- Muscle is held out with an Allis forceps and cut free from the surrounding muscle on either side of the cylinder of muscle.
- It is most important that the muscle sample is not squeezed or damaged in any way during this procedure, so that artifact changes are avoided.
- Finally, the specimens are transported in a vial (procured beforehand from neuropath lab) rapidly to the laboratory [2].

Post muscle biopsy advice:

- Suture removal after 7 days.
- Analgesics and antibiotics for 5 days.
- Alternate day dressing of the wound.
- Keep the wound site dry and clean.

Complications:

- Bleeding
- Hematoma formation
- Wound infection
- Scar formation after healing [2]

Skin Biopsy

Indications:

- Less invasive diagnostic evaluation for various muscular dystrophies including dystrophinopathies and limb girdle muscular dystrophies.
- Evaluation of Progressive Myoclonic Epilepsy phenotype- such as Neuronal Ceroid Lipofuscinosis or Lafora body disease.
- Evaluation of Neuronal Brain Iron Accumulation [3].

Prerequisites:

- A short admission for daycare should be made.
- Skin biopsy can be done by giving local anesthesia, but younger children may sometimes require sedation for performing the procedure. Ensure that the child is fasting for at least 4 hours before the procedure in case of infants 3 hours of fasting is enough. However, if they need to wait further IVF should be started.
- Proper, written and informed consent should be taken from the parents. If parents are not available then

whoever has brought the child for treatment needs to give consent on behalf of the child parents. The parents and guardians should be explained about the procedure, risk of sedation /local anesthetic use and possible complications and consent form should be signed.

- The vial for transport of skin biopsy sample duly obtained beforehand from the neuropathology laboratory [3]
- Ensure that in the treatment room sedation is ready Midazolam and Ketamine in labeled syringes with appropriate dilution, Lignocaine, sterile set, blade, suture.

Steps:

- The skin biopsy site is identified e.g., axilla
- The skin around the biopsy site is shaved and disinfected.
- A local anesthetic is administered into the skin and subcutaneous tissue⁴.
- Punch biopsy is done using skin biopsy needle.
- External pressure is applied with the free hand which will minimize the bleeding.
- Sterile dressing has to be applied
- Biopsied skin sample has to be transported in formalin, glutaraldehyde vials for routine/electron microscopy to the Neuropathology lab.
- If skin biopsy is performed in a child along with muscle biopsy, skin biopsy sample can be taken from the site of incision of muscle biopsy using the sterile blade / scalpel [4].

Post skin biopsy advice:

- Alternate day dressing of the wound.
- Keep the wound site dry and clean.

Nerve Biopsy

Indications:

- Inflammatory neuropathies – vasculitis, sarcoidosis, leprosy;
- Genetic neuropathies – HNPP, giant axonal neuropathy
- Metabolic disorders, with distinctive features and storage inclusions (sometimes only visible at the ultrastructural level) e.g. metachromatic leukodystrophy, Krabbe's disease, Fabry's disease, polyglucosan body disease;
- Tumour infiltration
- Toxic neuropathies, with characteristic changes, e.g. amiodarone, solvent abuse [5].

Contraindications

- On anticoagulant/antiplatelet agents
- Risk/benefit needs to be evaluated when there is peripheral vascular disease, edema.

Prerequisites:

- A short admission for daycare should be made.
- Ensure that the child is fasting for at least 4 hours before the procedure in case of infants 3 hours of fasting is enough. However, if they need to wait further IVF should be started.
- Proper, written and informed consent should be taken from the parents. If parents are not available then whoever has brought the child for treatment needs to give consent on behalf of the child parents. The parents and guardians should be explained about the procedure, risk of sedation /local anesthetic use and possible complications and consent form should be signed.
- The vial for transport of nerve biopsy sample duly obtained before hand from the neuropathology laboratory.
- Ensure that in the treatment room sedation is ready Midazolam and Ketamine in labeled syringes with appropriate dilution, Lignocaine, sterile set, blade, suture.
- Oxygen tubing, mask, flow meter, suction catheter should be kept ready in treatment room.

Procedure

- Patient is placed in prone position with the ankle slightly everted so that the foot is at a right angle to the leg. Part cleaned and draped.⁶
- For local anesthesia 0.5% lignocaine without adrenaline may be infiltrated from the lateral malleolus to around 10 cm proximal to it. A longitudinal skin incision is made midway between the lateral malleolus and posterior border of ankle around 4-5 cm proximal in the trough lateral to the Achilles tendon.
- The lesser saphenous vein if visible/palpable acts as an excellent landmark for the skin incision. The subcutaneous tissue is dissected out beneath which the semitransparent, tough Scarpa's fascia is visible.
- After the fascia is incised open, the saphenous vein can be identified adjacent to which the sural nerve lies. The lesser saphenous vein can be made prominent by compressing the calf⁶.
- The nerve branches are placed at an acute angle whereas the tributaries of the vein are at a right angle. Care should be taken not to stretch the nerve.
- The nerve's glistening white appearance with a pattern of alternating longitudinal lighter and darker bands helps distinguish it from veins and other tissues.
- The nerve can be dissected out from the surrounding tissue and with the help of suture the 2 ends of the nerve should be tied and a 3-5 cm's of nerve has to be cut by a sharp scalpel/blade.
- Biopsied nerve should be placed on saline moistened gauze and transferred to the vials obtained and

transported to the lab immediately (Neuropathology). The skin is sutured back and the site bandaged.

Monitoring during /after procedure:

Continuous vital parameter monitoring is necessary during the procedure particularly when IV sedation is given. Patient needs to be closely observed till sensorium is regained. The patient wound should be observed for 4-6 hours to monitor for any delayed wound site bleeding/swelling [6].

Post nerve biopsy advice:

- Analgesics and antibiotics for 5 days.
- Alternate day dressing of the wound.
- Keep the wound site dry and clean.
- Suture removal after 7 days.

Complications:

- Bleeding
- Hematoma formation
- Wound infection
- Wound dehiscence and neuroma formation
- Scar formation after healing

- Focal neurological deficit [6]

Conclusion

Nerve, muscle and skin biopsy are essential procedures required for establishing diagnosis for various neuromuscular and neurodegenerative disorders. Timely performing these procedures, where there is appropriate clinical conditions can help in early diagnosis and timely management. However, proper sterility and postoperative care is empirical to avoid complications.

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