# Spontaneous massive haemothorax in the peri-partum period of an undiagnosed neurofibromatosis type 1 patient - A surgical perspective

**Narasimman Sathiamurthy, MMed Surg[1], Hari Dass Govindasamy, MD[1], Kanesh Kumaran Seevalingam, MD[2], Umasangar Ramasamy, MD[2], Ganendra Paramasvaran, MD[2]**

[1]Thoracic Surgery Unit, Department of Surgery, Hospital Sultan Abdul Halim, Sg Petani, Kedah, Malaysia, [2]Department of Surgery, Hospital Taiping, Perak, Malaysia

**SUMMARY**

Acute massive haemothorax is a life-threatening situation, which is often associated with a preceding trauma. However, spontaneous haemothorax is a rare occurrence, especially in pregnancy. Spontaneous haemothorax in the immediate post-partum period secondary to a ruptured intercostal AVM is extremely rare more so in the background of an undiagnosed neurofibromatosis. This is a report of a young lady presenting with pleuritis and breathlessness after the delivery of her 1st child. Her management is discussed.

**INTRODUCTION**

Acute massive haemothorax is a life-threatening situation, which is often associated with a preceding trauma. However, spontaneous haemothorax is a rare occurrence, especially in pregnancy. Spontaneous haemothorax in the immediate post-partum period secondary to a ruptured intercostal AVM is extremely rare more so in the background of an undiagnosed neurofibromatosis.[2] This is a report of a young lady presenting with pleuritis and breathlessness after the delivery of her 1st child. Her management is discussed.

**CLINICAL SUMMARY**

Miss M, a 33-year-old woman at day-4 of post-partum, with no significant past medical history, presented with sudden onset of right sided pleuritic chest pain associated with shortness of breath. She had no previous history of trauma, surgical intervention, or rib fracture. Her perinatal period was uneventful.

There was a hyperpigmented skin lesion measuring 5x7cm over the right side of the posterior chest wall (Fig 2A). A subtle scoliosis was observed. Breath sounds were absent on the right side with dullness on percussion. The chest radiograph showed a massive right-sided pleural effusion and a dorsal scoliosis. A diagnostic pleural tapping in the emergency department yielded frank blood. She remained hemodynamically stable and contrast enhanced CT thorax was performed, which revealed a sinister lesion appearing as a possible right intrathoracic intercostal AVM (arterio-venous malformation) with no active contrast extravasation at the time of study (Fig 1). A chest drain was inserted into her right hemi-thorax and 1800cc of dark blood was drained. She was nursed in the High Dependency Unit (HDU), while

arrangements were being made for a transfer to a tertiary centre for percutaneous angio-embolisation. The patient suddenly became hypotensive in Stage II shock. Decision was made for an emergency thoracotomy to arrest the bleed.

In the operating room, a right lateral thoracotomy was performed. The thoracic cavity was filled with blood clots and there were two distinctive AVM arising from the posterior chest wall over the 7th intercostal space measuring 5x5cm and 4x3cm respectively. The bigger AVM had an ulcerated wall with active ooze of blood (Fig 2B). Haemostasis was achieved by ligating and excising the AVM. Post operatively, she was still dependent on vasopressors. A repeat CT angiogram to evaluate for residual bleeding showed no evidence of active contrast extravasation, however incidentally noted a hematoma extending into her spinal canal, causing compression of the cord at T7/T8 level. MRI of the spine was done and confirmed these findings and uncovered few other lesions suggestive of a peripheral nerve sheath tumour along the T5/T6 and T6/T7 with cord compression only at T7/T8 level. Subsequent clinical examination revealed a myelopathic type of neurological deficit over her right lower limb with a preserved anal tone. She was transferred to a neurosurgical facility where she underwent an emergency decompressive laminectomy of segment T7/T8 and evacuation of clots.

Post-laminectomy, she was stable and extubated. Unfortunately, she became paraplegic with loss of bladder and sphincter functions. She underwent intensive rehabilitation post operatively, however her neurology remained the same. She was discharged home after a month in the hospital. At six-month post-surgery, there were no changes in her neurological status.

The histopathology result of the excised AVM showed mesenchymal proliferative lesions, which tested positive for S100 and consistent with neurofibroma plexiforme without atypia.

*This article was accepted: 18 March 2018*
*Corresponding Author: Narasimman Sathiamurthy*
*Email: drnara@hotmail.com*

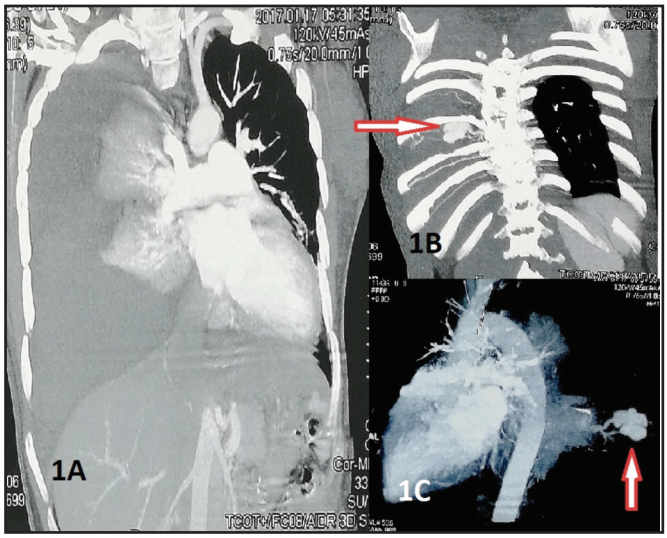


**Fig. 1:** A) shows the extensive right hemothorax causing mediastinal shift and right lung collapse.
B) shows the location of the AVM at the 7th intercostals space.
C) demonstrates the feeding vessels to the AVM.

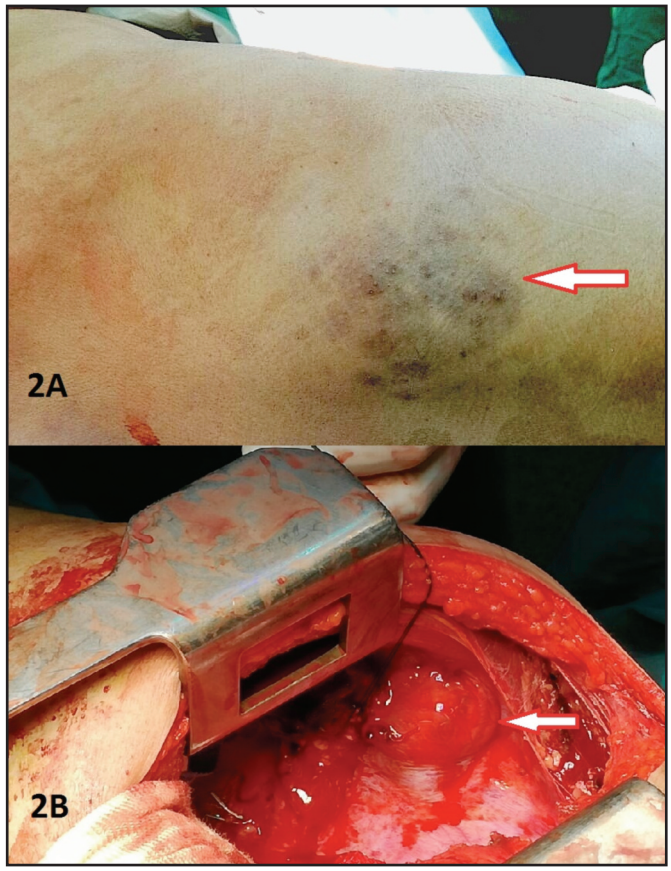


**Fig. 2:** A) shows the hyperpigmentation of the right side of posterior chest wall with café-au lait patches.
B) is the intra-operative photograph of the ruptured right intercostal AVM.

## DISCUSSION

Common causes of respiratory distress in pregnancy include pulmonary embolism, asthma, amniotic fluid embolism and pulmonary oedema. Spontaneous haemothorax however is a rare cause and occurs without any inciting factors.[2,5]

Causes of spontaneous haemothorax in pregnancy could be separated into vasculopathic and non-vasculopathic causes. Non-vasculopathic causes include ectopic pregnancy, gestational trophoblastic disease, ruptured pleural adhesions and vasculopathic causes include pulmonary arteriovenous malformations (PAVM), Hereditary Haemorrhagic Telangiectasia (HHT) or Osler-WeberRendu Syndrome (OWR), dissection or rupture of arterial aneurysms, Type IV Ehlers-Danlos Syndrome, and neurofibromatosis (NFM).[1] In our case, ruptured intercostal AVM in pregnancy is an extremely rare in the background of undiagnosed neurofibromatosis with only a few case reports available in the literature.[2]

Diagnosis is often established late due to low index of suspicion leading to significant morbidity and mortality, as seen in Miss M[2,5]. Presenting symptoms are highly variable and may include abdominal pain, dyspnoea, thoraco-abdominal masses, flank pain, and/or shoulder and back pain.[1]

Pregnancy accounts for 50% of ruptured AVM in women under the age of 40, largely attributed to the hemodynamic and hormonal changes that occur during pregnancy.[1] Increase in intrathoracic pressure, blood volume and cardiac output will increase the blood pressure, which will lead to dilatation and rupture of AVM. Elevated steroid levels in pregnancy increase vascular fragility. Risk of dissection or rupture is more prominent in the third trimester and in the immediate post-partum period.[1]

The histopathology evaluation of the tissue sample obtained intra-operatively in our patient was reported as neurofibroma.

Neurofibromatosis type 1 (NF1) is a hereditary autosomal dominant disorder originating in the neuroectoderm and mesoderm. It is expressed in approximately 80% of those who have the gene, with incidence of about 1:3000.[4] NF1 is characterised by abnormal cutaneous pigmentation and multiple skin tumours, and it is frequently associated with scoliosis and other skeletal abnormalities. The incidence of vascular lesions has been reported to be only 3.6%.[3,4] However, the frequency of blood vessel involvement in NF1 has been underestimated, primarily because lesions may be clinically silent. Pregnancy is thought to exacerbate neurofibromatosis if present. In addition to the present case,

the literature contains less than 30 cases of massive intrathoracic haemorrhage in patients with NF1.[1-3] It is also been thought previously that the presence of neurofibromatosis in itself renders the blood vessels fragile with or without aneurysm.[4]

Miss M is a patient of undiagnosed NF1 evident by presence of hyperpigmentation, café au lait patches and scoliosis, whom unfortunately suffered a sub-clinical AVM that became more susceptible to the increased intrathoracic pressure and hormonal changes leading to its rupture during her peripartum period. Due to the presence of lesions likely to be neurofibroma along her spinal canal and the proximity of the AVM to the intervertebral foramina, the blood clot could have tracked into the spinal canal from the AVM site and caused a mass effect leading to the post-operative paraplegia. The frequent location of these peripheral nerve sheath lesions near the spinal canal could pose a challenge in achieving haemostasis while preventing secondary mass effect.

## CONCLUSION

Spontaneous haemothorax due to ruptured intercostal AVM during peripartum period is an extremely rare life-threatening emergency. Physiologic changes of pregnancy influencing the intrathoracic pressure, cardiovascular and endocrine system contribute significantly to the fragility of the AVM. The clinical signs such as hyperpigmentation and café au lait patch with scoliosis in Miss M should have given the clue of possible underlying AVM in the peripartum period and options of further investigations could have been discussed. Angioembolisation is preferred prior to surgical intervention whenever permissible. In the event of massive haemothorax with circulatory collapse of an immediate post-partum lady, the anxiety to arrest the bleeder may lead to undesired complications and care must be taken to minimise collateral damage.

## REFERENCES


1. Doshi HM, Robinson S, Chalhoub T, Jack S, Denison A, Gibson G. Massive spontaneous hemothorax during the immediate post-partum period. Tex Heart Ins J 2009; 36(3): 247-9.
2. Parashi HS, Bhosle KN, Thakare ND, Sharma A, Potwar SS. Giant congenital intercostal arteriovenous malformation with extensive involvement of chest wall and ribs: surgical experience. Ann Thorac Surg 2013; 95(6): 2157-9.
3. Oderich GS, Sullivan TM, Bower TC, Gloviczki P, Miller DV, Babovic-Vuksanovic D, et al. Vascular abnormalities in patients with neurofibromatosis syndrome type I: Clinical spectrum, management, and results. J Vasc Surg 2007; 46(3): 475-84.
4. Von Recklinghausen F. Uber die multiplen fibrome der Haut and ihre Beziehung zu den multiplen Neuromen. Berlin: A. Hirschwald; 1882. p. 138.
5. Chen CY, Chen CP, Wang KG, Kuo SC, Su TH. Factors implicated in the outcome of pregnancies complicated by acute respiratory failure. J Reprod Med 2003; 48(8): 641-8.