INTRODUCTION
Sickle cell anemia is an autosomal recessive genetically transmitted hemoglobinopathy responsible for considerable morbidity and mortality. This is a hereditary disorder caused due to defective hemoglobin structure. In 1952 it was recorded for first time simultaneously amongst the tribal population group of nilgiri hill and laborers in the tea garden of Aasam. Now it is firmly established that these genes harbor amongst different caste groups but with very high prevalence amongst scheduled castes, scheduled tribes and other backward communities of India. According to studies carried out by Kate, 10% of total population of the state of Maharashtra belongs to the tribal population groups. In Maharashtra, Banker et al reported prevalence of the disease from 1.9% to 33.5% in different communities. This disease has variable clinical presentation and most of the Indian patient remains asymptomatic for longer periods due to higher levels of HbF. Sicklecell disease presenting as death in clinically asymptomatic patient with sickle cell disease or sickle cell trait is not uncommon. Here, we are presenting two case reports that are of great medico-legal importance and challenge to the clinician as well as autopsy surgeon.

Case report 1: 27yrs old married female of scheduled tribe community of north Maharashtra region who was primi-gravida, with 38wks of gestation was admitted to general (civil) hospital Dhule. No antenatal treatment record/ history was available with the patient. She was provisionally diagnosed as a case of pre-eclampsia and anemia. Emergency lower segment caesarian section (LSCS) was done on next day under spinal anesthesia and she delivered a live baby girl. Operation was uneventful. After the operation patient developed sudden breathlessness, chest pain, pain in extremities and died within five hrs of operation. Following her death, allegations of medical negligence were raised by her husband and relatives. So after police submitted the inquest papers, medico-legal post-mortem examination was carried out by team of six doctors from forensic medicine, pathology, obstetrics and gynecology, and anesthesia. On external post-mortem examination, she was found to be moderately built. Edema was present over feet and lower legs. Evidence of (intact sutured wound) of surgical intervention in the form of LSCS was noted in her lower abdomen. Apart from this no other external injury was noted. Cyanosis was absent. On internal post-mortem examination, brain & lungs were congested & edematous. Lungs were spongy and cut section showed froth. Thoracic cavity contained 120ml of serous fluid. Trachea and laryngeal cartilages were intact. Heart was

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normal in size and shape with all valves, chambers & coronaries normal. Hematoma was present over rectus abdominis muscle in mid-line of abdomen, reddish in color, in the area of 15cm x 15 cm, weighing 350gms. Peritoneal cavity contained 300gms of blood and blood clots. Liver (1300gms) spleen (110gms) & both kidneys (rt 90gms, lt 80gms) were intact. On cut section of kidneys, cortico-medullary distinction could be appreciated and renal capsule could be easily stripped off. Uterus was showing intact stiches of surgical intervention of LSCS. Signs of pregnancy and recent delivery were present. Uterine cavity contained minimal blood clots. Stomach was intact, empty and having no peculiar/ abnormal smell. All abdominal organs were pale. Organs were preserved for histopathological examination and chemical analysis. Chemical analysis report of viscera didn’t reveal any poison in the exhibits. Histo-pathological report reveal sickled RBCs in lungs, liver, spleen, kidneys, brain. Kidneys showed acute tubular necrosis. Histo-pathology examination of piece of rectus muscle showed hemorrhagic areas with sickled RBCs. Histo-pathological examination of endo-metrial surface of uterus showed decidua and areas of hemorrhage. The hemorrhagic areas showed sickled RBCs. Myometrium were hypertrophied. Both ovaries and fallopian tubes showed sickled RBCs. 

So based upon clinical presentation of the patient, post-mortem findings, chemical analysis report and histo-pathological findings, we gave our final opinion as to the cause of death as “Sickle cell crisis (vaso-occlusive crisis) associated with pre-eclampsia and anemia”, in a woman with undiagnosed sickle cell trait.

Case report No 2: A male prisoner of 43yrs age, of scheduled tribe community of north Maharashtra region who was lodged in central jail, complained of chest pain, while he was in his barrack. He was immediately brought to the hospital in the jail premises where he was declared dead on admission. Relatives of the dead complained of torture by the prison authorities. Medico-legal post-mortem examination was conducted at SBH Govt medical college, Dhule by panel of doctors comprising of forensic medicine & pathology. On external examination of dead body, there were no injuries over the dead body. On internal
examination of dead body, no injuries were noted. Brain (wt 1250gm) was congested; both lungs (right 550gms & left 500gms) were congested & oedematous. Pleural effusion was present in left thoracic cavity, 50cc yellowish in color. Heart was enlarged, fatty deposition was present around heart and petechial hemorrhages were present on posterior surface. Left ventricular thickness was 2cm & anterolateral wall showed whitish fibrotic areas. Liver (wt 1100gms), spleen (wt130gms) & both kidneys (wt150gms each) were congested. Stomach was empty having no peculiar smell, mucosa of stomach was intact. Viscera were preserved for chemical analysis & histo-pathological examination.

Chemical analysis report of viscera didn’t reveal any poison in the exhibits. Histo-pathological report revealed sickled RBCs in lungs, liver, spleen, kidneys, brain. In lungs pulmonary oedema was noted. Section from heart showed atherosclerotic changes with lumen narrowing in left anterior descending coronary artery. Section from whitish fibrotic areas showed myocardocytes replaced by fibro-collagenous scar, suggestive of healed myocardial infarct. Congested blood vessels showing sickled RBCs. Pericardium showed hemorrhagic areas.

So based upon clinical presentation of the patient, post-mortem findings, chemical analysis report and histo-pathological findings, we gave our final opinion as to the cause of death as “Sickle cell crisis (vaso-occlusive crisis) associated with coronary artery disease”, in a man with undiagnosed sickle cell trait.

**DISCUSSION**

The clinical features of sickle cell anemia result more from the vaso-occlusive consequences of sickle cells than from the anemia itself. The term sickle cell crisis was introduced to describe a recurring attack of pain involving the skeleton, chest, abdomen or all three. Using the term in a broader sense, vaso-occlusive, “crises” comprised a variety of syndromes that are typically recurrent and potentially catastrophic. Clinical manifestations are sudden in onset and are directly attributable to obstruction of the micro-circulation by intravascular sickling. Typically, the dorsa of both the hands and feet are swollen, non-erythematous and painful. In adults a triggering event is not often identified. A perceived precipitating factor includes skin cooling, emotional stress, physical exertion and pregnancy\[^{5,6}\]. Although rare, exertional collapse and sudden death are the most serious potential complications of sickle cell trait due to sickle cell crisis. Dehydration, hypoxia, acidosis and physical exertion are known aggravating factors for sudden death in sickle cell trait due to vaso-occlusive cell crisis\[^7\].

The factor precipitating sickle cell crisis in first case may be pregnancy or anxiety disorder associated with labour or LSCS. Despite the medical advances in recent decades, pregnancy is still associated with many clinical and obstetrics complication in patient with sickle cell disease resulting in a higher maternal mortality.

Oxygen demand during pregnancy increases to support the metabolic requirements of the placenta and fetus. As the maternal oxygen reserve may be comprised during pregnancy due to the increased oxygen consumption and decreased functional residual capacity, patient may be predisposed to hypoxemia with exacerbation of sickling and its complications. These changes during pregnancy highlight need for a multi-disciplinary team of experts to monitor pregnant sickle cell women in a tertiary hospital\[^7\].

Similarly as seen in case report 2, the absence of external and internal injuries excluded death due to assault. The absence of petechial hemorrhages in the conjunctivae and the absence of injuries in the neck exclude an asphyxial death due to neck compression or strangulation\[^8\]. There has been increased awareness & investigations of sudden death in police custody, prison. Not all deaths occurring during restraint of a person are necessarily unnatural but forensic pathologist should be careful while dealing with such cases and viscera should be preserved for chemical analysis & histo-pathological examination for arriving correct cause of death\[^8\].

The factor precipitating sickle cell crisis in this case may be emotional stress associated with imprisonment. In both the cases histo-pathological section of all organs showed disseminated intravascular sickling suggestive of vaso-occlusive crisis leading to multi-organ failure and death.

Dr R V Bardale et al noted that 5% of pregnant women who died with sickle cell disease had past mediastinal history of the disease and in 4.76% cases the cause of death was sickle cell disease\[^9\].
Ana Cristina Silva- Pinto et al reported that 4% of pregnant women who were suffering from sickle cell disease had pre-eclampsia while 2.9% of maternal deaths were due to sickle cell crisis\(^7\).

**CONCLUSION**

Clinical profile of sickle cell patient in north Maharashtra region is less severe as compared to African countries and characterized by late onset of presentation, more a symptomatic presentation and lower mortality. Hence, most of the patient remained undiagnosed. So it is not uncommon to see such cases presenting as deaths without any significant history. In this case the patient presented without any significant present or past symptoms of sickle cell disease.

In first case report, medical negligence leading to death was ruled out because of meticulous autopsy done by autopsy surgeons. In the second case report, custodial torture/ unnatural death was ruled out by careful autopsy. Therefore the autopsy surgeons should be aware about sickle cell crisis as a cause of death. There has been increased awareness & investigations of sudden death in police custody/prison. Not all deaths occurring during restraint of a person are necessarily unnatural but forensic pathologist should be careful while dealing with such cases. Hence proper autopsy, clinical history, gross findings in the organs and careful histopathological examination is necessary.

To avoid maternal mortality, we suggest that, each and every pregnant lady of scheduled tribes, scheduled castes, and other backward communities should be carefully screened for sickle cell trait in the first trimester of pregnancy to avoid complications/ loss of life to both mother as well as baby. Affected pregnant woman should be looked after by a unit experienced in the care of women with this condition. Blood transfusion may be needed in some women with poor obstetric history or a severe form of sickle cell disease. Regular folic acid, prompt treatment of infections and crisis and increased fluid intake make it possible for most women to have a successful outcome of pregnancy\(^5\).

**REFERENCES**

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