Continuing Importance of Autopsy in Neurodegenerative Diseases

During the past several decades, advances in imaging and other diagnostic tools have suggested the demise of the autopsy as a useful procedure. Additionally, there is little understanding of the importance of autopsy not only among the lay public but also among many clinicians due to the truncation of pathology teaching in many medical school curricula. The object of this article is to correct some of these misperceptions about the autopsy.

What goes on in an autopsy?

The autopsy begins with a detailed inspection and measurement of the external features of the body. For a complete autopsy, an incision is made in the chest and abdomen through which the internal body organs are removed and examined. Grossly observable abnormalities are carefully noted and appropriate portions of the various organs are retained for further studies. Even in the evaluation of central nervous system diseases, studies of other organs are important since renal disease, liver disease, cancer, hypertensive cardiovascular disease, and other disorders can contribute to or be primarily responsible for a patient's neurological illness.

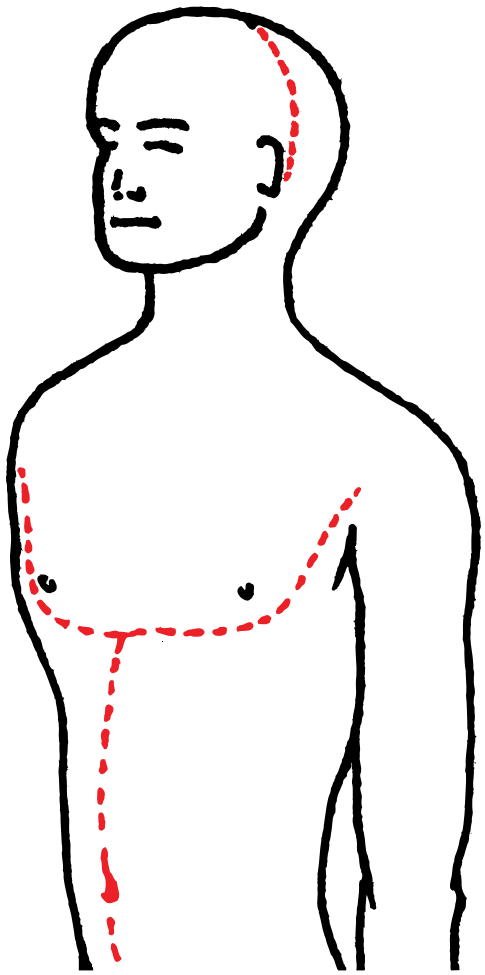

Figure. Drawing of the human upper body illustrating the position of incisions (dotted lines) made during an autopsy. The Y-shaped incision made in the body exposes the organs of the chest and abdomen; the incision over the top of the head from ear to ear is made to remove the brain. For viewing at the funeral, the body incision is covered by clothing and the head incision is hidden by the pillow in the casket

The brain is exposed by incising the scalp from ear to ear and sawing the skull circumferentially to produce a skull cap that is lifted off to expose the meninges, blood vessels, and cranial nerves. After the brain is removed, the skull cap is replaced and the scalp is folded back together and carefully sutured. The removed brain can then be studied to identify the disease process.

Value of the Autopsy Brain Material

The complex internal structure of the brain, with marked anatomical and biochemical variations from one area to another, makes its analysis different from the study of more homogeneous organs, such as the liver or spleen. Unfortunately, computerized images closely resembling the gross appearance of the brain at autopsy have provided physicians a false sense of security regarding the accuracy of such techniques for understanding (during life) various disease processes of the brain. Too many clinicians believe that they already know everything of importance about a patient's central nervous system from these images, and therefore feel there is no need for an autopsy to "confirm" these "findings." In fact, only relatively gross changes in the brain are identifiable with imaging techniques. Moreover, studies that can only be performed on autopsy brain tissue (enzyme histochemistry, receptor labeling, immunocytochemistry, and DNA and RNA analysis) provide important information on the fine details of the disease process.

Currently, there are no diagnostic tools other than histopathologic study of the brain with which to establish accurate diagnoses of the progressive degenerative diseases of the nervous system. Brain biopsy is associated with an unacceptably high rate of morbidity or mortality and provides tissue from only a limited region of the brain; brain biopsy also does not reveal the extent of a disease and can miss focal changes. Therefore, the pathological examination of autopsy brain remains the only means of conclusively establishing the diagnosis of Alzheimer's disease, Pick's disease, Creutzfeldt-Jakob disease, idiopathic Parkinson's disease, and many other neurodegenerative diseases. In addition to establishing definitive neuropathologic diagnoses, the brain autopsy material is crucial for providing investigators with clues to the underlying causes of these neurodegenerative disorders.

Despite the fact that most neurodegenerative disorders affect older adults, there has been a marked decline in the rate of autopsies performed on older individuals: up to 80 percent of deaths of persons in their thirties are evaluated by autopsies, as compared to only about 5 percent of the deaths of persons over age eighty years. One reason for such a low autopsy rate in elderly individuals is that most patients with neurodegenerative diseases die at home or in a chronic care facility. Therefore, arrangements that allow autopsies to be performed on individuals who die outside of a hospital are particularly important for this population. Most professional funeral directors are willing, usually at only a small additional charge, to transport a body to a hospital at which preparations have been made to perform the autopsy.

Clinicians must recognize the value of the information that can be obtained from an autopsy and must play an active role in obtaining permission from families. The request for autopsy permission should be carried out in an empathic manner, and clergy and social workers who are known to the family can also be of help. Advocacy groups, such as the Alzheimer's Association through its Autopsy Assistance Network, have helped increase the lay public's awareness of the importance of the autopsy for understanding neurodegenerative disorders. Facts about the autopsy must be provided to the families of deceased individuals in order to dispel the myths that surround this procedure. Even religious beliefs need not preclude the performance of an autopsy, since a family's resistance may be based on misinformation about religious tenets concerning the dissection of bodies. The mistaken idea that an autopsy "mutilates" the body so that funeral directors cannot embalm the body or prepare it for viewing must be dispelled. Finally, and perhaps most important, physicians must recognize that beyond the "confirmation of clinical impressions," the autopsy still is the only method for making an unequivocal diagnosis of many neurodegenerative diseases, and that it is also the only means of providing tissues for investigative efforts directed at understanding, preventing, or curing these disorders. ✦

REFERENCES:

1. Further details can be obtained from the book chapter: Brumback RA, Eskin TA, Lapham LW, Panner BJ: The autopsy as an important diagnostic and research tool in neuroscience. In Mayeux R, Gurland B, Barrett VW, Kutscher AH, Cote L, Putter ZH (eds): Alzheimer's Disease and Related Disorders: Psychosocial Issues for the Patient, Family, Staff, and Community (Charles C. Thomas: Springfield, IL, 1988) pp. 135-143.

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